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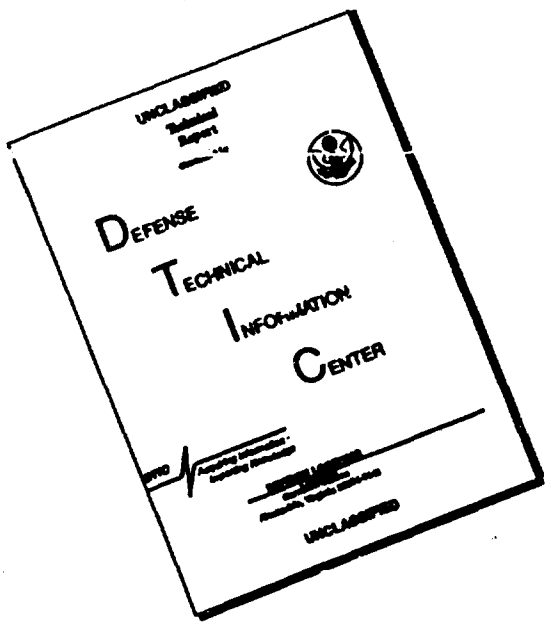

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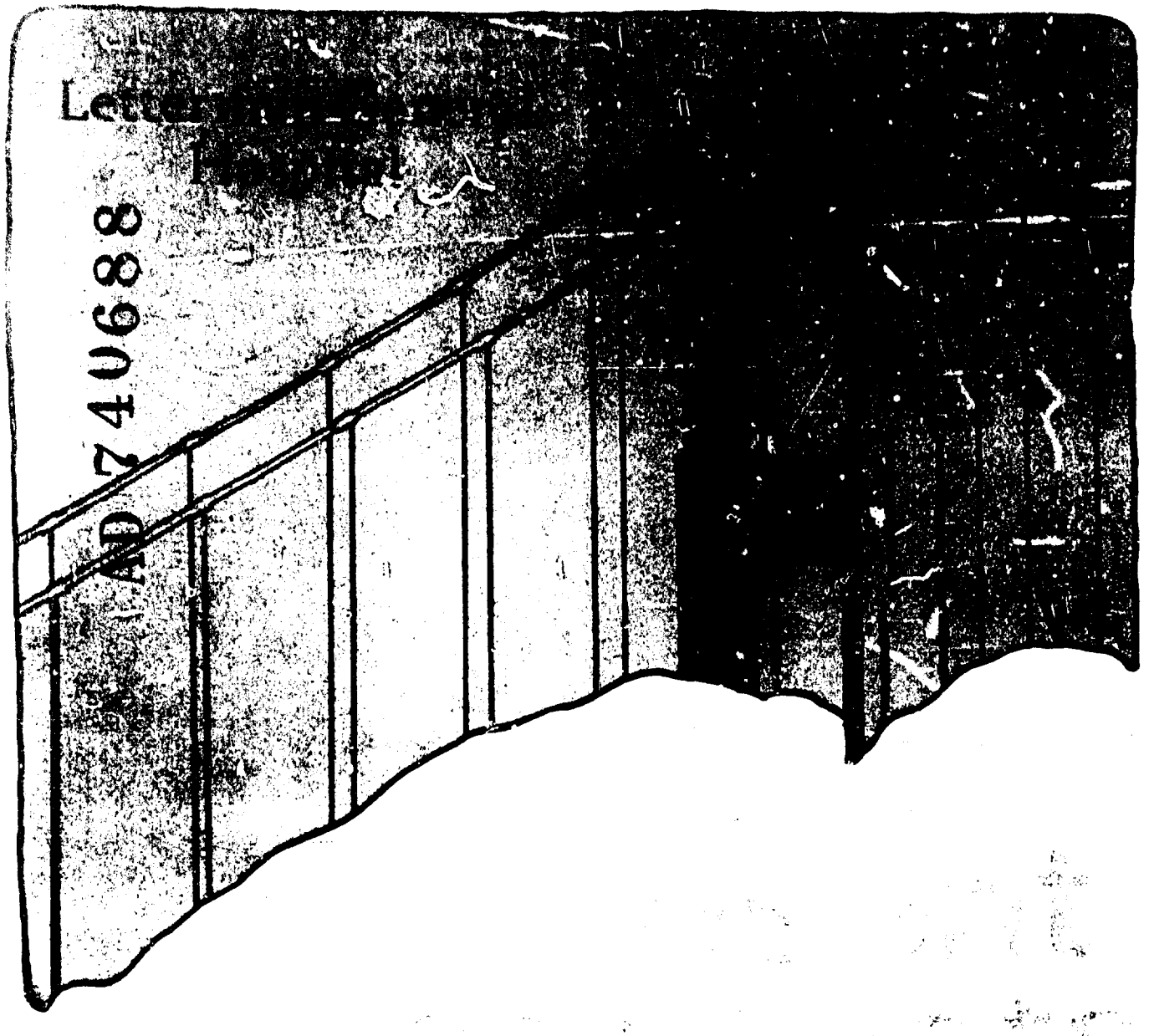

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13. ABSTRACT

This ten-article symposium presents some of the more unusual and complicated medical problems of children. Actual cases cared for by the pediatricians at Letterman General Hospital are reported and discussed and appropriate references provided for further study. Many of the diseases presented are not new to physicians; however, only in recent years has some understanding and methods of treatment become available. For example, the literature has come forth with studies on the "host defense mechanism". The first article in this symposium describes an infant with a severe lack of host defenses. It can also be recognized that infants may point also to disease in the mother. A report of two infants with hypocalcemia shortly after birth is illustrative that their problems were secondary to a previously unsuspected disease in the mother. The article on the infant of the diabetic mother demonstrates the importance of teamwork -- the mother, the obstetrician, the internist, the pediatrician -- so as to insure a viable infant who can then develop normally. Following this paper is a report of four illustrative cases on heart disease in the newborn infant. Only in recent years has attention been given to congenital heart disease, not because of lack of interest, but the physicians' having to await the technical advances necessary for catheterization and cardiac surgery in infants. Recognizing cyanosis, heart failure, and respiratory distress, and then knowing when and what to do are the pediatrician's first steps in saving these infants' lives. The author provides for the physician workable instructions parallelling the case reports and additional discussion for background.

The fifth paper describes the complicated care required of the typical low-birth weight infant. The next article reports four children with infantile spasms followed

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by an article with a description of Reye's syndrome (and case report) This pictures the sequence of events which terminates in coma and hepatic failure. There is a report of a case of purulent constrictive pericarditis in an infant who did survive without surgical intervention.

Because better preventive medicine in this country is available, complications of tuberculosis are rarely seen in children today; however, these still are problems in some medically undeveloped countries. The report of a two-year-old Thai girl with multiple pseudocystic tuberculosis of the bone is an example not only of the disease but reflects one experience a young Army physician had during his residency to become acquainted with world health problems.

The final article reviews the Letterman General Hospital experience in cases of child abuse November 1970-November 1971. A series of tables present the data, and the author describes not only the "hypothetical child" but the "typical parent" who commits child abuse. Comments by the author on the problem as a "social disease which must be attacked at each of its roots" are complemented by his remarks on the contructive team approach (Infant and Child Protective Council) which meets weekly to report the independent research and to develop solutions to the problems.

14. KEY WORDS	LINK A		LINK B		LINK C	
	ROLE	WT	ROLE	WT	ROLE	WT
PEDIATRICS						
IMMUNOLOGIC DEFICIENCY SYNDROMES						
HYPOCACEMIC TETANY						
DIABETES, Infant of Diabetic Mother						
Infant, low birth weight						
SPASMS, infantile						
REYE'S SYNDROME						
PERICARDITIS, constrictive in infants						
TUBERCULOSIS, multiple pseudocystic						
BONE, tuberculous						
CHILD ABUSE						

PRESENT CONCEPTS

IN INTERNAL MEDICINE

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Guest Editor

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LETTERMAN GENERAL HOSPITAL
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December 1971

PRESENT CONCEPTS IN INTERNAL MEDICINE
VOLUME IV *December 1971* Number 12

PEDIATRICS
SYMPOSIUM

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FORTHCOMING SYMPOSIUM. . .

GASTROENTEROLOGY

(The Small Bowel)

Present Concepts, Vol IV No 12, December 1971

FOREWORD

The *alpha* and *omega* . . .

In the first issue of this volume of *Present Concepts in Internal Medicine* we presented some of the common everyday child health care problems. In the final issue of this volume we present some of the more unusual and complicated medical problems of children. Actual cases cared for at Letterman General Hospital are presented and discussed with appropriate references provided for further study. Many of the diseases presented are not new to physicians; however, only in recent years has some understanding and methods of treatment become available. Greater understanding is the result of technical advances and of specializing by more physicians in the problems of children.

Most of our knowledge of the host defense mechanism has developed since 1952 when colonel Ogden Bruton, then chief of the Pediatric Service, Walter Reed General Hospital, described the first child with agammaglobulinemia. It is significant to note that he was not satisfied with only treating the infections in the child but wanted to know the reason why the child was having so many infections. In order to live in this world, the human organism is dependent on its defense mechanism against other organisms. The first article describes an infant with a severe lack of host defenses.

Infants can also point to disease in the mother. Doctor Pfuetze describes two infants with hypocalcemia shortly after birth. Their problem was secondary to a previously unsuspected disease in the mother.

Caring for the infant of the diabetic mother demonstrates the importance of team work. The mother, the obstetrician, the internist and the pediatrician must work together to insure a viable infant who can then develop normally. The problems of hypocalcemia, hyaline membrane disease, hypoglycemia and hyperbilirubinemia are discussed by Doctor Robinson.

Only in recent years has attention been given to heart disease in the newborn. This has not been due to lack of interest but has awaited the technical developments of safe cardiac catheterization and cardiac surgery in infants. Recognizing cyanosis, heart failure, and respiratory distress in an infant and knowing what to do are the pediatrician's first steps in saving these infants' lives. Doctor Farina demonstrates the expertise and knowledge brought to this subject.

Foreword

Doctors Robinson and Done describe the complicated care of an infant who developed infection, apnea, hyperglycemia, hyperbilirubinemia, and hypocalcemia, and presented a feeding problem. More nurses adequately trained in the care of the sick newborn infant, neonatologists, and well-equipped intensive care nurseries can improve the infant mortality rate in the United States. The case presented illustrates the multiple considerations that are necessary and therapy which was effective for this infant.

Convulsive disorders in children are frightening to parents, children, and physicians. Early diagnosis and treatment of infantile spasms is important to prevent mental retardation. Reports on four such children are presented by Doctor Epstein.

A syndrome of coma and hepatic failure known as Reye's syndrome has been observed more frequently recently. Adequate therapy requires knowledge of cause. It is perplexing to see a child die and not understand why. Doctor Epstein describes the disease course and therapeutic attempts.

It has been rare for an infant to survive with constrictive pericarditis. Doctor Weir describes a case of purulent constrictive pericarditis in an infant who did survive.

During a three months rotation in the Children's Hospital, Bangkok, Thailand, Doctor Weir observed a child with complications of tuberculosis. Due to better preventive medicine in this country, these complications are rarely seen. Doctor Weir was able to gain a better knowledge of world health problems while in Thailand.

There have always been cases of child abuse. Only in the past ten years have pediatricians become more involved in protecting the lives of children. Under the direction of Doctor Reister, the Infant and Child Protective Council has been established at Letterman General Hospital and meets each Friday. Doctor Reister reviews our experience during the period November 1970 to November 1971. It is of utmost importance that the infant and child being abused be identified and protected against further abuse, and that the child abuse be recognized as a social disease which must be attacked at each of its roots.

Children are the nation's most valuable resource. We must recognize their problems and help them develop to be adults who can serve themselves and others to their best ability.

LTC James L. Stewart, Jr., MC
Guest Editor

GRAFT VERSUS HOST REACTION IN AN IMMUNOLOGIC INCOMPETENT CHILD

LTC James L. Stewart, Jr., MC

In 1952 Bruton observed an absence of serum gammaglobulins was associated with repeated bacterial infections in an eight year-old boy at Walter Reed General Hospital. Since that observation many investigators have attempted to unravel the complicated mechanisms that a host uses to resist infectious agents. This complex interaction between host and infective agent is only partly defined.

Largely based on experimental evidence produced by Cooper and Good at the University of Minnesota, a two component concept of immunity has immerged: (1) a cellular immune system based on the lymphocyte and (2) a humoral system based on the plasma cell. Other mechanisms contribute, such as, the complement system, macrophages, polymorphonuclear leukocytes, and interferon.

The following case report concerns an infant with dual system defect -- failure in cellular immunity and humoral immunity. Possibly this is an example also of intrauterine onset of graft versus host reaction.

CASE REPORT

On 12 February, a girl, weighing 9 lbs, was born after a normal 40 weeks of gestation, to a 24-year old primagravida. The mother had received chlorothiazide and vitamins during pregnancy. At birth the infant had a generalized erythematous papulosquamous skin rash over the entire body which was diagnosed as possible Leiner's disease. The rash improved with adrenocortical steroid cream applications and became worse when steroids were discontinued. For three weeks she was breast fed and then received a soy bean formula. At four weeks of age, skin care consisted of Alpha-Keri®, Neutragena® soap and Valisone® cream. The rash cleared.

On 24 March she was admitted to the hospital because of diarrhea and pneumonia. Treatment consisted of penicillin, kanamycin and intravenous fluids. She improved and was discharged on 6 April. Ten days later (nine weeks of age) she was admitted to the hospital for a nine day stay because of fever, diarrhea, and dehydration. The hematocrit was 34 percent; hemoglobin 10.8 gm; white blood cells (WBC) 17,300/cu mm with a differential count of 63 percent neutrophils, 21 percent bands, 11 percent lymphocytes, and 4 percent monocytes. The serum sodium was 150 mEq/liter, potassium = 4 mEq/liter, chloride = 113 mEq/liter, carbon dioxide = 18 mEq/liter. Spinal fluid was normal. Fecal and nose cultures revealed Staphylococcus aureus. Improvement was obtained with intravenous fluids, ampicillin, neomycin, Isomil®formula and rice cereal.

She was again admitted to the hospital on 4 May and weighed only 7 lb 11 oz because of dehydration. A fecal culture grew Staphylococcus aureus and a pathogenic Escherichia coli. Serum immunoglobulins were as follows: IgG 113 mg/100 ml, IgA 32 mg/100 ml, IgM 70 mg/100 ml. Gantrisin® was given for a urinary tract infection. The skin rash was worsening and the diarrhea was chronic. Formula had frequently been changed involving Similac®, Isomil® and Pro-Sobee®.

She was transferred to Letterman General Hospital on 18 May at three months of age, weighing 8 lb 12 oz. Length was 21 inches. The pulse rate was 144 beats/min. She followed a light with her eyes. She was extremely irritable. The body was covered with a dry, flaky, desquamating rash. A greenish exudate was present around the ears, neck, and inguinal areas. The cheeks were puffy. Decreased muscle mass was noted. The eyelids were swollen and red. No mucosal lesions were present. Auscultation of the chest was normal. The liver was palpable 4.5 cm below the right costal margin. The spleen was not palpable. The puffiness of the cheeks was thought to be edema of hypoalbuminemia. Four to eight watery yellow bowel movements occurred each day. Laboratory data on this admission are noted in Table 1.

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TABLE 1

LABORATORY DATA. Immunologically Incompetent Infant.*

SERUM Constituents				Protein Electrophoresis		Immunoglobulins	
	mg/100 ml		*mEq/liter*		*gm/100 ml*		*mg/100 ml*
Calcium	8.1	Sodium	141.0	Total protein	3.0	IgG	130
Glucose	140.0	Potassium	4.2	Albumin	1.5	IgA	24
Blood urea nitrogen (BUN)	15.0	Chloride	106.0	$Alpha_1$ globulin	0.3	IgM	0
		Carbon		$Alpha_2$ globulin	0.7		
Phosphorous	3.3	dioxide	25.0	Beta globulin	0.3		
Cholesterol	75.0			Gamma globulin	0.1		
Triglycerides	90.0			OTHER			
				Alkaline phosphatase 195 units/100 ml LDH 255 Wacker units SGOT 55 Karmen units			

HEMATOCRIT	RETICULOCYTES	WHITE BLOOD CELLS† Total - 17,100/cu mm		LYMPHOCYTES	
% per volume	*%*	*% per volume*		*%*	
25	2.3	Neutrophils	59	Small	36
		Bands	4	Intermediate	56
		Metamyelocytes	8	Large	12
		Myelocytes	6		
		Eosinophils	8		
		Monocytes	2		

PROTHROMBIN TIME %	FECES	BACTERIOLOGIC CULTURES Site	Organism
75	Fat = negative x 4 Reducing substance = negative x 2 pH 6.0 and 5.0	Skin	Candida species, *Staphylococcus aureus*, coagulase positive, Pseudomonas
		Feces	*Staph. aureus, Escherichia coli* type 026:B6
		Throat	*Staph. aureus*
		Mouth	Candida species
		Blood	*Staph. aureus*, Pseudomonas species
		Urine	negative

*Fourteen weeks old. May 1971, LGH

†Leukocyte nitroblue tetrazolium test = normal

Roentgenograms of the chest, skull, and long bones were considered normal. A skin biopsy revealed hyperkeratosis, parakeratosis, increased histiocytes, and an absent granular layer. The liver biopsy showed moderate fatty metamorphosis with non-caseating granulomas. Bone marrow aspiration showed myeloid hyperplasia. Sixty-three percent granulocytes (including six percent eosinophils), 17 percent lymphocytes, 10 percent reticulum cells, 10 percent erythroid elements, and no plasma cells were found. The possibility of Letterer-Siewe disease or Runting disease was considered.

After consultation with the Dermatology Service a diagnosis of congenital icthyosiform erythroderma was suggested and treatment with three percent lactic acid cream to soften the scabs and the application of Burrow's solution soaks to the weeping areas was advised.

Antibiotic therapy consisted of oral neomycin, systemic Coly-Mycin® for pseudomonas and oxacillin for staphylococcus. A triglyceride formula (Portagen®) was given as oral feeding. Whole blood, 100 ml of compatible A positive, was given on 22 May. Marked improvement in weight, bowel movements, activity and skin rash occurred.

She was discharged on 13 June 1970 to continue Portagen® formula, ferrous sulfate, multivitamins at home. It was evident that an immune deficiency disease was the basic cause manifested by hypogammaglobulinemia. It was advised that no immunizations be given. In July she was treated for otitis media with erythromycin and oxacillin. The tip of the spleen was palpable at that time.

On 4 September the infant, weighing 12 lb 8 oz, was again admitted to Letterman General Hospital. Skin rash had persisted with varying degrees of acuteness. Chronic diarrhea was present. The skin lesions were patchy, erythematous, and in some areas, raised and scaly. The finger nails and toe nails were poorly formed and had broken ends. The hair was fine and sparse. The hematocrit was 41

percent; WBC 17,300/cu mm with 50 percent neutrophils, 25 percent lymphocytes, 18 percent eosinophils, one percent monocyte. Diagnosis of left upper lobe and right middle lobe pneumonia was made and treated with penicillin. She was discharged on 5 September to be followed in the Pediatric Hematology Clinic of the University of California Medical Center, San Francisco.

On 8 September she was admitted to the University of California Medical Center, San Francisco, because of left lower lobe collapse. The lung disease continued to progress. The diagnosis of Pneumocystis carinii infection was considered. Because of probable histiocytosis, therapy of prednisone and Velban® was started. A progressive worsening led to death.

Postmortem Microscopic Examination

Lungs. Alveolar spaces were filled with granular debris consistent with Pneumocystis carinii which were definitely identified by Gomori's methenamine silver stain. Thymus. The small thymus was grossly abnormal. Corticomedullary differentiation was lacking. No Hassall's corpuscles were found. The thymus consisted only of epithelial and stromal elements. Lymph nodes. Peripheral lymphoid tissue lacked both lymphocytes and plasma cells in both thymic dependent and independent regions. No germinal centers were present. There was an increase in eosinophils and histiocytic appearing cells.

DISCUSSION

Deficiencies of lymphoid tissue which carry out humoral and cell mediated processes result in susceptibility to recurrent infections. The case presented represents deficiency in thymic-dependent or cell-mediated immunity and in thymic-independent immunity or humoral immunity.

Congenital agammaglobulinemia was first described in 1952 by Bruton. Two types of inheritance are known, the X-linked recessive disorder observed only in males and the autosomal recessive type affecting males and females. Infants are typically well until nine months of age when an undue susceptibility to encapsulated pathogens such as pneumococcus, Hemophilus influenzae,

staphylococcus, meningococcus, and pseudomonas is manifest. This is probably because of an inability of the polymorphonuclear leukocytes to ingest and digest these organisms without the participation of opsonins. The response to viral and fungal infections is usually normal. A syndrome of diarrhea, steatorrhea, and an increased frequency of autoimmune disorders is common. Diagnosis is made by immunochemical determinations of serum gamma globulins. Antibodies are absent. Circulating lymphocytes are normal. The delayed allergic response is normal. Homograft immunity is normal. The thymus gland shows well-populated deep cortical areas but a deficiency in cells and germinal centers in the far cortical areas. The absence of germinal centers and plasma cells in lymph nodes following stimulation of lymph nodes establishes the diagnosis. Treatment consists of gamma globulin injections 0.7-1.0 ml/kg, intramuscularly, every 2-4 weeks. The disease is to be distinguished from physiologic agammaglobulinemia in infancy acquired agammaglobulinemia secondary to lymphoproliferative diseases, nephrosis, and exudative enteropathy. Other antibody deficiency syndromes are shown in Table 2. Many of these are referred to as dysgammaglobulinemias.

TABLE 2

OTHER ANTIBODY DEFICIENCY SYNDROMES
Transient Antibody Deficiency of Infancy

Antibody deficiency with . . .
splenomegaly
elevated IgM
normal IgG
decreased IgM
thyoma
nodular lymphoid hyperplasia of intestine
steatorrhea
pernicious anemia
Antibody deficiency without . . .
immunoglobulin deficiency
Deficiency of IgA

A defect in the cellular immunity system, thymic dependent, represents a greater threat to life than an isolated fault of the humoral system. Cellular immunity is manifested by the demonstration of a delayed hypersensitivity skin test reaction

independent of the presence or absence of circulating antibodies. Studies show that a bone marrow stem cell is modulated by the thymus in early life and takes up residence in defined areas of lymph nodes and spleen. Specific antigens sensitize these cells by means of interaction with macrophages so that a number of immune factors are elaborated on contact later with the same antigen. These factors activate a population of mononuclear cells which are capable of mounting an assault on the infecting organism. The delayed hypersensitivity reactions are important in the defense against such viruses as rubeola, varicella, and vaccinia; fungi such as monilia; and tuberculosis.

Two syndromes have been recognized associated with the thymus gland and cell mediated immunity. DiGeorge syndrome (congenital aplasia of the thymus) is associated with the absence of the thymus gland and parathyroid glands. There does not appear to be a hereditary basis. Infants with this defect suffer from hypocalcemia and a propensity to viral and fungal infections. Lymph nodes have normal thymic independent germinal centers and medullary cords with depletion of deep cortical thymic dependent areas. There is an inability to manifest cell mediated response; however, serum immunoglobulins and antibody responses are normal. These patients are candidates for thymic grafts.

Nezelof's syndrome (thymic dysplasia with normal immunoglobulins) has faulty development of the thymus gland possibly associated with a failure of bone marrow stem cells to migrate to the thymus gland. The thymus gland lacks corticomedullary differentiation and Hassall's corpuscles, remains small, and fails to descend into the mediastinum. Severe and recurrent infections of candidiasis, varicella, Pneumocystis carinii, disseminated vaccinia following smallpox are frequently lethal early in infancy. Normal serum immunoglobulins are present. Impaired cell-mediated immunity is present. Transplants of normal thymic tissue may prove helpful.

These two syndromes in humans illustrate the counterpart in experimentally thymectamized animals which shows deficiencies of cell mediated immunity and relatively intact humoral immunity.

Combined deficiencies involving both cell mediated (thymic dependent) and humoral (thymic independent) immunity are the most serious immunologic defects known. Earlier reports were made by investigators in Switzerland and the term "Swiss type agammaglobulinemia" is often used. Several forms of thymic

dysplasia and variable deficiencies of immunoglobulins are now recognized. Some are transmitted as X-linked recessive and others are autosomal recessive. An early onset of bacterial, viral, and fungal infections plague the infant. Progressive respiratory disease, intractable diarrhea, and persistent candida infections are manifest. Disastrous results are obtained with immunizations with live virus vaccines especially smallpox and BCG. Serum immunoglobulins are abnormal. There is an absence of delayed hypersensitivity reactions and defective homograft rejection. The thymus is rudimentary and has no Hassall's corpuscles. Peripheral lymph nodes lack lymphocytes and plasma cells and thymic-dependent and thymic-independent regions. These children are prone to graft versus host reactions. Whole blood transfusions contain viable lymphocytes which can initiate a graft versus host reaction. A chronic form of graft versus host disease has been recognized due to transfer of maternal cells into an immunologically incompetent host in utero. A desquamated, erythematous maculopapular eruption occurs with alopecia of the scalp and eyebrows in the chronic form of graft versus host. Defects of cell-mediated immunity not associated with obvious thymic abnormalities have been reported.

The infant described in this report demonstrated abnormally low serum immunoglobulins. Adequate test for delayed hypersensitivity deficiency were not performed because of the skin rash; however, the autopsy findings showing a markedly abnormal thymus gland and peripheral lymph nodes demonstrated a deficiency in the thymic dependent lymphocytes. These findings indicate a deficiency in both the humoral immunity and cell-mediated immunity in this infant. Doctor Robert Good reviewed the autopsy findings and thought there was also evidence of a chronic graft versus host reaction initiated in utero by maternal lymphocytes in an immunologic deficient fetus.*

In treating this type of patient it is easy enough to replace the gamma globulin with standard gammaglobulin preparations containing predominantly IgG. This therapy does not aid the cellular immune deficiency. Attempts to replace the missing cells have been made. Results are disappointing. The transfer of cells is difficult. The half-life of most cells is short. The specificity of immunocompetent cells will produce a graft versus host reaction. Some bone marrow cell transplants have been successful if the graft versus host reaction is prevented.

*Personal communication.

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As for myself, I take the ground, that when a patient applies to a physician he applies with the view of being cured, and cares not what may be the means used, so that they are not of an injurious character–hence, I employ whatever means I believe capable of effecting a cure, without regard to the source from which they are derived–and if I can perform the cure solely through mental influence and placebo remedies, I will not adopt any other course–it is my privilege and right as a physician to do so–and it is a duty which my patient expects and requires of me. His grand object is to be cured–mine, to cure, without a slavish deference to books or authorities. The Almighty has given to every man brains, mind, reasoning powers, judgment, etc., for his own use and to determine his own actions in life; and he, who dare not employ his mind and act for himself, but who tamely yields to the dictates of others, is unfit to be a freeman–is indeed, already the most contemptible of all slaves.

–JOHN KING, M.D.*

*From *Chronic Diseases*, a book authored by King. Although the publisher is unknown (title page was missing from the volume I have), I believe this is the work of the famous John King, M.D., from South Carolina who practiced medicine in the nineteenth century in the lower part of the state. This volume, I am told, was printed about 1850. The portion quoted appears on page 84.

JLS

NEONATAL HYPOCALCEMIC TETANY
Secondary to Maternal Hyperparathyroidism

MAJ Bruce L. Pfuetze, MC, and LTC James L. Stewart, Jr., MC

Neonatal tetany is most often attributed to either the high phosphate content in cow's milk or "first day" hypocalcemia./1/ Hypocalcemic tetany in the newborn associated with parathyroid overactivity in the mother appears to be rare. Friderichsen /2/ reported the first case in 1938. This article concerns two hypocalcemic infants with irritability and seizures whose mothers had serum calcium and phosphorus studies consistent with hyperparathyroidism. Surgical exploration post partum revealed parathyroid adenomas in both mothers.

CASE REPORTS

CASE 1. A 16-day old, Caucasian male was admitted to Letterman General Hospital for evaluation of seizures. The infant was delivered as a breech presentation after a 38-week gestational period which was uncomplicated, except for premature rupture of the membranes. At 24 hours of age, the infant began regurgitating small amounts of blood and passed black tarry material, rectally, on two occasions. The infant was treated with ampicillin and kanamycin for five days for suspected septicemia; however, all blood cultures were negative. On the eighth day of life, the infant was discharged in apparently good health.

From the tenth to the sixteenth days of life, the parents noted that the infant was extremely jittery and had intermittent jerking of the head and arms. These spells lasted about 20 seconds and were followed by a sleep period. The infant was otherwise doing well and took 120 ml of Similac® every 4 hours.

Initial physical examination was normal except for the neurologic findings. The infant was extremely tremulous and had frequent seizures manifested by first right-sided jerking of the extremities with the eyes rolling back, followed by generalized seizure activity lasting approximately 20 seconds. Reflexes were hyperactive with unsustained clonus, bilaterally.

Initial laboratory studies were as follows: white blood cells 13,400/cu mm with 46 percent neutrophiles, 43 percent lymphocytes, eight percent monocytes, three percent eosinophiles; hematocrit 38 percent; serum CO_2 21 mEq/liter; chloride 104 mEq/liter; sodium 135 mEq/liter; potassium 4.3 mEq/liter; venous pCO_2 30 mm Hg; venous pH 7.45; serum blood urea nitrogen 9 mg/100 ml; glucose 90 mg/100 ml; calcium 9.2 mg/100 ml. Lumbar puncture revealed clear fluid with no cells, sugar 56 mg/100 ml and protein 75 mg/100 ml. Feces studied for occult blood were negative. Cultures of the spinal fluid, blood, urine, and feces revealed no significant bacterial growth. Bone age, chest and skull roentgenograms were within normal limits. Echoencephalogram was normal. The electroencephalogram was abnormal, characterized by paroxysmal sharp wave activity which was bisynchronous but with slight left-sided preponderance.

Phenobarbital (6.0 mg intramuscularly) was given twice daily and increased to 8 mg intramuscularly, every 8 hours. On the twenty-second day of life, the infant was started on ampicillin because one blood culture was positive for anerobic streptococcus. While on phenobarbital the infant was free of seizures but remained jittery and had unsustained clonus. At 24 days of age, a serum calcium was 6.5 mg/100 ml and phosphorus was 7.2 mg/100 ml. Calcium gluconate 0.5 gm was added to each 120 ml of Similac® to provide an additional 2.0 gm of calcium gluconate daily. During the next several days, clonus and jitteriness decreased. At 26 days of age, repeat calcium was 8.2 mg/100 ml, phosphorus 4.8 mg/100 ml, total protein 5.7 gm/100 ml, magnesium was 0.9 mg/100 ml (normal serum

magnesium 1.4± 0.2). Because the infant was still shaky with slight clonus, he was given 0.8 ml of 50 percent magnesium sulfate ($MgSO_4$), intramuscularly, and then 500 mg (1.0 ml of 50 percent $MgSO_4$) in each 120 ml of Similac®. At the time of discharge, the patient continued to have slight jitteriness but the clonus had disappeared. He was discharged on iron-enriched Similac® with 10 ml of 10 percent calcium gluconate and 0.5 ml of 50 percent $MgSO_4$ added to each 120 ml of formula.

Ten years previously, the 30-year-old mother had poliomyelitis with cranial and spinal nerve involvement and four spontaneous pneumothoraces. She had had one kidney infection four years previously and a bladder infection during her last pregnancy, but she had no history of hematuria or passage of renal calculi. For the past several years, she has had intermittent pain in her knees, wrist and sternum. She has noted that she tires easily and has been irritable during the past several months. Two other children, ages 7 and 3 years, are in good health. The mother's family history was unremarkable. Physical examination was essentially normal, except for a slight bulge of the right lobe of the thyroid. Scintiscan and radioactive iodine uptake (RAI) study of the thyroid were normal. The serum calcium was 12.8 mg/100 ml and phosphorus was 1.3 mg/100 ml. Repeat studies revealed calcium 12.7 mg/100 ml and phosphorus 2.1 mg/100 ml. At surgical exploration of the neck, a 4.0 cm parathyroid adenoma was removed.

CASE 2. A 10-day old Oriental female was admitted to Letterman General Hospital for "shaking spells" involving the left side of the face, the tongue and the left hand which the mother had noted about four times daily for two days. On the day of admission, the infant had had seven seizures, each lasting 15-30 seconds. This patient was the 3331 gm (7 lb 5 oz) product of a full-term gestation in a 23-year-old primagravida. Pregnancy,

labor and delivery were uneventful, except for forceps rotation from right occipital transverse to occiput anterior. The neonatal course was uneventful and the infant was discharged on the third day. The infant took 90 ml of Enfamil® every 3 to 4 hours and was in good health except for seizures. There was no history of trauma. The infant's mother and father were both in good health and there was no other familial history of seizures.

Physical examination at the time of admission revealed an alert, active infant whose vital signs were: temperature 98.6 F (37 C) rectally; pulse 124/min; respirations 46/min; length 48 cm; weight 3969 gm (8 lb 8 oz), and head circumference 37 cm. Examination was unremarkable except for hyperactive reflexes with unsustained clonus and Chvostek's sign. An observed seizure lasting 30 seconds consisted of rhythmical twitching of the left side of the face, tongue, arm and leg.

Initial laboratory studies included: blood sugar 100 mg/100 ml; white blood cells 10,900/cu mm with neutrophiles 30 percent, lymphocytes 60 percent, and hematocrit 31 percent. Spinal fluid had 800 red blood cells/cu mm and one lymphocyte/cu mm. Phenobarbital (20 mg intramuscularly) was given and repeated one hour later. Seizures continued and 2.0 ml of 10 percent calcium gluconate was given intravenously but the patient continued to have seizures. Paraldehyde 0.6 ml in 10 ml of 5 percent dextrose in water was given over 15-20 minutes. Following this dose of paraldehyde, the seizures occurred about every 30 minutes. Approximately two hours later, the infant began having seizures more frequently and was given 0.6 ml of paraldehyde in 10 ml of 5 percent dextrose in water over 30 minutes. At this time, serum calcium was reported to be 5.6 mg/100 ml and the phosphorus was 11.9 mg/100 ml. After 6 ml of 10 percent calcium gluconate had been given slowly to the infant, the seizures stopped. Calcium gluconate (500 mg/kg/day) was added to the infant's formula, and no further seizure activity was noted.

The mother's serum calcium was 12.0 mg/100 ml and the phosphorus was 2.3 mg/100 ml. She was in good health with no specific complaints. She denied tiredness, paresthesias, bone pain, and passage of urinary calculi. She described one episode of flank pain and hematuria in 1969. There was no abnormality palpable in the neck. Repeated laboratory studies revealed calcium 12.0 mg/100 ml, phosphorus 2.2 mg/100 ml. Alkaline phosphatase was 24 King-Armstrong (KA) units (normal = 4-17 KA units). Barium swallow, thyroid scan, abdominal and hand roentgenograms were normal. A 1.5 cm parathyroid adenoma was removed at surgery.

COMMENT

The two cases reported illustrate neonatal hypocalcemic tetany secondary to maternal hyperparathyroidism. Van Arsel /3/ has suggested that hyperparathyroidism is not conducive to pregnancy. Walton /4/ reported an increased incidence of stillbirth. In 1962 Ludwig /5/ reviewed the complications of pregnancy and of fetal morbidity and mortality in a survey of women with hyperparathyroidism and noted an increased incidence in this population.

The mechanism of intrauterine suppression of the fetal parathyroid gland is controversial. The suppression is probably due to an excess of maternal calcium or parathormone. Whether or not parathyroid hormone crosses the placenta is still questioned. It also appears that suppression may not be mediated by calcium ions because suppression of the fetal parathyroid has been observed in offspring of both hypocalcemic and hypercalcemic mothers./5/

Hypomagnesemia observed in the first infant may have contributed to the seizures./6/ This possibility should be considered and treated appropriately.

Our two cases point out the significant influence of maternal hyperparathyroidism contributing to symptomatic

hypocalcemia manifested by seizures in the offspring. When tetany is observed and the infant is hypocalcemic, appropriate therapeutic measures for the infant should be employed, and one should evaluate the mother for hyperparathyroidism.

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THE INFANT OF THE DIABETIC MOTHER

CPT Carl D. Robinson, MC

Farquhar /1/ in 1959 described the infants of the diabetic mother as "remarkable not only because like fetal versions of Shadrach, Meshach and Abednego, they emerge at least alive from within the fiery metabolic furnace of diabetes mellitus, but because they resemble one another so closely that they might well be related." The infant of the diabetic mother (IDM) has one of the few pathologic conditions that generally can be diagnosed by its appearance and can alert the obstetrician for the future management of the mother, and the pediatrician in the immediate management of the offspring.

CASE REPORT

The infant weighing 5 lb 13 oz, was delivered by Caesarean section at 37 weeks gestation to a 22-year-old gravida II, para I, abortion I, class C diabetic. (Table 1). The mother has had diabetes since age 14 and has required 40 units NPH per day. She was hospitalized three weeks prior to delivery for control of diabetes which required an increased insulin dose to 50 units NPH per day. Her weight gain was 16 lb. At birth the infant had Apgar of 5/7. Respiration was immediate but the heart rate initially was slow. The infant was plethoric and cushinoid in appearance. It exhibited within the first hour marked expiration grunting and labored respiration. Chest x-ray revealed a reticular granular pattern which was consistent with hyaline membrane disease. A umbilical cord blood sugar was 300 mg/100 ml; however, a blood sugar at one hour of age was 40 mg/100 ml. A catheter was inserted, appropriate fluid administered, and blood gases were monitored. At 24 hours of age, the infant required assisted ventilation; it was placed in an infant respirator and given high oxygen

concentration. It was weaned from this system by six days of age. On day 3, the bilirubin began to rise and peaked on day 6 at 14.6 mg/100 ml. Phototherapy was used. It was also noted that the serum calcium during the third and sixth days was between 5.2 and 6.5 mg/100 ml. The infant remained asymptomatic and no intravenous calcium was given. With time and maintenance of acid base balance, the calcium rose to normal by the eighth day. The child was discharged home on the 15th day. Followup through the year and at one year of age revealed essentially a developmentally normal one-year-old girl.

DISCUSSION

The incidence of maternal overt diabetes is approximately 1:116-1:200, whereas gestational diabetes occurs 1:500-1:1000. /2/ This is a disease of high risk to both the mother and the fetus. The major problems the obstetrician has to face with these mothers are toxemia, hydramnios, urinary tract infection, and pyelonephritis. The neonatal morbidity and mortality are significantly different between the offspring of the diabetic and nondiabetic mothers. The major causes of morbidity and mortality of the infant with a diabetic mother are hypoglycemia (approximately 50 percent /3/), hyaline membrane disease (has also approached 50 percent in past years /4/, congenital anomalies (most commonly skeletal and heart disease, three times normal incidence /5/), and birth trauma /6/. In addition, these infants may have hypocalcemia (25 percent) and hyperbilirubinemia (25 percent), and an increased incidence of congestive heart failure, hyperkalemia, renal vein thrombosis, and infection. In general, the perinatal pediatric mortality is influenced by the severity of the diabetes as judged by vascular complications./7/ Infant mortality ranges from 0.0 to 4.8 percent in Class A to 28.0-47.8 percent in Class F. (Table 1). Good obstetrical prenatal care and careful medical management in controlling maternal diabetes is highly significant in reducing infant mortality and morbidity.

TABLE 1

WHITE'S CLASSIFICATION (MODIFIED). A METHOD TO HELP CATEGORIZE THE RISK TO THE INFANT OF THE DIABETIC MOTHER (IDM)

CLASS	DESCRIPTION DIABETES IN MOTHER
A	Gestational diabetes, prediabetes
B	Diabetes after age 20 without vascular disease. Diabetes for less than 10 years duration.
C	Diabetes of greater than 10 years duration and with minimal vascular disease.
D	Diabetes with onset before age 10 with evidence of vascular disease including retinitis, albuminuria, and hypertension.
E	All of D plus calcification of pelvic vessels.
F	All of D plus nephritis.

Management of the IDM

The treatment of the IDM may be divided into two phases (a) prenatal, where the treatment of the mother effects the development of the fetus, and (b) postnatal, when the infant is treated directly.

During pregnancy and at the time of delivery, the goal is to maintain normoglycemia in the mother. The method of management of the pregnant diabetic patient, in general, depends on the severity of her diabetes. The Class A diabetic is managed as nearly as possible as the nondiabetic patient while Classes B-F require multispecialty involvement of the obstetrician, internist, and pediatrician. These patients, because of the increased risk to the fetus and themselves are hospitalized for any deviation from the normal. Strict attention is paid to control of hyperglycemia, glucosuria, diet, and prevention of other maternal complications. Usually the infant is delivered between the 35th and 37th gestational weeks (although the timing may vary from hospital to hospital). The time of delivery is determined by how well one is able to control the maternal disease and how the fetus is tolerating its environment.

The pediatrician's primary role at the birth of the IDM is to maintain homeostasis in the infant. One must approach these infants with a high degree of suspicion and with some knowledge of the potential problems one can face. Hypoglycemia (defined as a blood sugar less than 30 mg/100 ml in the term infant and 20 mg/100 ml in the pre-term infant), if symptomatic, is treated with intravenous glucose and if the infant proves resistant to this therapy, glucagon, and ACTH have been used./8/ Hyaline membrane disease has been treated with parenteral fluids, blood, oxygen, buffers, and respiratory support as necessary. Because in most institutions the IDMs are delivered between the 35th and 37th weeks, the incidence of hyaline membrane disease has been high. Now with Glucks' sphingomyelin lecithin ratio /9/ which apparently has an extremely high predictability of lung maturity, we have a better index as to the possible occurrence of hyaline membrane disease and the feasibility of maintaining the pregnancy until maturity is assured. Hypocalcemia, if symptomatic, is treated with appropriate replacement of calcium gluconate and maintenance of proper acid-base balance. The reason for the occurrence of hypocalcemia and tetany is unknown, but it is certainly related to pH, serum protein, and the ionized calcium fraction. Hyperbilirubinemia may be treated with phototherapy /10/ and, if necessary, with exchange transfusion.

Our case study report points out the complex problems the IDM faces after the first breath. For this patient these complications included prematurity, hyaline membrane disease, hyperbilirubinemia, and hypocalcemia. This infant appeared clinically to match the features of a IDM. Pathophysiologically, the findings that are characteristic include increased body fat, decreased extracellular water, relatively decreased total body water, visceromegaly, decreased muscle weight, and islet cell hyperplasia. /3,11/ Biochemically, the alterations in the serum insulin, pancreatic insulin, and tissue glycogen are elevated; and the blood glucose, serum free fatty acids, serum growth hormone, and catecholamine excretion are decreased./12/ At present, there is no unifying hypothesis that can account for the many biochemical derangements that occur in the infant of the diabetic mother.

SUMMARY

The pregnant diabetic and her offspring are among the most challenging problems the obstetric and pediatric team can face. It is a pathologic state where proper management of a maternal condition can improve markedly the fetal and neonatal outcome. It is also a disease of high risk to both mother and fetus. The question of why so many metabolic derangements occur to the infant of the diabetic mother has yet to be answered. At the present, one can anticipate and approach the problems as they clinically present in hopes of improving the overall prognosis of these infants.

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Probably just as true today –

> Do not insist on antipyretic measures in every fever; a moderate fever seems to be a part of the body's means of ridding itself of an infection. If the child seems fairly comfortable with a temperature of one hundred and three degrees, let it alone.*

*From *Golden Rules of Pediatrics*, John Tahorsky, A.B., M.D. C.V. Mosby Medical Book Co., St. Louis 1906.

CONGENITAL MALFORMATIONS OF THE HEART

Four Case Reports Illustrating Diagnostic and Therapeutic Application of Cardiac Catheterizations in Infants

MAJ Matthew A. Farina, MC

The incidence of congenital malformations of the heart is reported to be five in each 1000 live births. About one-third of all babies born alive with a congenital heart defect will die during the first month of life.

There are relatively few ways in which the infant with heart disease can present. Most commonly, the features are cyanosis, congestive heart failure, cardiac murmurs, abnormal heart rate, or a combination. The case reports selected for this paper represent four infants with different diagnoses and with different combinations of presenting features. Each had cardiac catheterization at Letterman General Hospital in the past twelve months. The diagnosis and distinguishing features of heart disease are given above each case history. Pertinent annotations appear in right hand column. A comment follows each case. Some of the comment relates to the specific diagnosis involved, but other parts of the comment provide general concepts which may be applied to any infant with heart disease.

CASE I

Cyanosis
Moderate systolic ejection murmer
Moderate right ventricular hypertrophy
Heart, normal size

Diagnosis: Tetralogy of Fallot

A 9-week-old infant was born after an uncomplicated pregnancy and had a birth weight 6 lb 11 oz. He was discharged at three days of age and did well at home until seven weeks of age when he was noted to become faintly cyanotic with crying. At eight weeks of age he had a deep cyanotic episode associated with tachypnea. He was admitted at US Army Hospital, Fort Ord, and was transferred to Letterman General Hospital.

Note the delayed appearance of cyanosis, even with significant heart disease. Consider, and rule out, neurologic disorders. Transfer such infants to appropriate facilities.

Physical examination revealed faint cyanosis at rest. All pulses were present and equal. Examination of the precordium revealed no abnormal pulsations. There was no abdominal organomegaly. On auscultation the first heart sound was normal, whereas the second heart sound was single. A long harsh 3/6 systolic ejection murmur was heard along the left sternal border.

The nature of the second heart sound is extremely important in Pediatrics.

Chest roentgenograms showed no cardiomegaly, but the heart was boot-shaped with a concave pulmonary artery segment. Vascularity was diminished. Electrocardiography demonstrated right axis deviation of 120 degrees, right atrial hypertrophy with peaked P waves, and right ventricular hypertrophy.

Consider diagnoses, Table 1.

On the evening of admission, the infant had two cyanotic episodes associated with tachypnea, precipitated by crying. During these spells the murmur virtually disappeared. The infant was initially treated with oxygen, 1.0 mg of morphine subcutaneously, and subsequently propranolol 4.0 mg three times a day orally. No spells were observed in the interval before surgery five days later.

Moniter such patients closely. This indicates that pulmonary blood flow is sharply decreased.

Emergency drugs and equipment should be at the bedside, in a Pediatric resuscitation cart.

Cardiac catheterization demonstrated tetralogy of Fallot with severe infundibular pulmonary stenosis. Two days later at open chest surgery, a Waterston anastomosis (right pulmonary artery to ascending aorta in a side-to-side relationship) was performed. Following this procedure a continuous murmur was heard in the second right intercostal space. The postoperative course was uneventful and the child was discharged. No cyanosis was present, and all medications had been discontinued.

COMMENT

Tetralogy of Fallot is the commonest form of cyanotic heart disease in most ages, but this is not true in the neonate and the infant in the first few months of life. When the malformation occurs, patients present with cyanosis or a loud murmur. The absence of a murmur in face of cyanosis in the newborn, is a poor sign. However, this diagnosis is straight-forward in a patient with slight to moderate cyanosis within weeks after birth, a moderate systolic ejection murmur, moderate right ventricular hypertrophy, and a normal sized heart.

In a clinical profile study, the hypoxic spells often seen in tetralogy of Fallot, are characterized by paroxysmal hyperpnea and increased cyanosis. The age of onset shows a peak incidence at one to three months of age. Spells occur primarily in the morning, usually related to awakening. The duration ranges from less than a minute to several hours; the majority last 15 to 60 minutes. Twenty-five percent of the patients have several spells per day. Crying has been noted to initiate the spells in 20 percent; feeding, defecation, exercise, and coughing, in descending order, also initiate spells. It has been observed that episodes usually do not follow severe exercise, but rather, occur in the morning after a good night's sleep. This suggests that a prolonged period of rest paradoxically heightens the susceptibility of the child to the paroxysms.

Hyperventilation increases cardiac output an average of 20 percent with no appreciable change in arterial pressure.

Therefore, systemic vascular resistance is reduced by approximately 20 percent. One of the proposed etiologies of paroxysmal hyperpnea is based on an oversensitivity of the respiratory drive mechanism. Paradoxically, this occurs after a period of rest. If prior to adjustment, a relatively sudden increase in activity or a Valsalva-like maneuver occurs (such as crying or defecation), the resulting increase in right-to-left shunting will immediately decrease arterial pO_2 and pH, and increase arterial pCO_2. This may trigger marked hyperpnea which in turn will increase cardiac output and decrease pulmonary blood flow, resulting in greater right-to-left shunting and greater arterial hypoxemia. A cyclic mechanism is established which may terminate in death.

In the past, morphine (1.0 mg/10 lb body weight) has been used to treat these attacks. The drug relaxes the restless, irritable child. It has been observed in tetralogy of Fallot that the degree of shunting may be considerably increased by drugs such as norepinephrine. This observation suggests that right outflow tract obstruction is increased by sympathetic nerve activity. Beta-adrenergic receptor blockage prevents the augmented response to sympathetic nerve stimulation, thus decreasing right ventricular outflow tract obstruction. Additionally, the increase in peripheral arterial resistance leads to a reduction in the right-to-left shunt, and an increase in pulmonary blood flow.

An intravenous infusion of propranolol in a dose of 0.2 mg/kg is recommended in the management of such attacks. Oral propranolol in doses of 1.0 to 4.0 mg every eight hours has been reported to be considerably less effective. In our patient it would be difficult to say whether the oral medication was effective or the attacks ceased spontaneously.

The major indications for a shunt procedure in tetralogy of Fallot are hypoxic spells, failure to thrive and gain weight, and significant cyanosis and polycythemia. The hypoxic spells usually cease after surgery. The Waterston procedure has become popular in infants because the procedure can be performed through a midline incision with relative ease. In the subsequent total repair, the anastomosis can be easily taken down as opposed to the Pott's anastomosis (descending aorta to left pulmonary artery). In the infant, the Blalock-Taussig anastomosis (subclavian artery to pulmonary artery) is often technically more difficult because of size of the subclavian artery.

CASE II

Congestive heart failure
Right ventricular hypertrophy
Increased pulmonary vascularity
Cardiomegaly
Murmer, soft systolic ejection

Diagnosis: Double outlet, right ventricle

A three week old infant was the first born child of a mother in Seoul, Korea. The antenatal course was benign, and birth weight was 7 lb 8 oz. Cyanosis was present at birth and did not clear despite the use of oxygen. There was no respiratory distress. No murmurs were heard and there was no evidence of heart failure. Chest roentgenograms revealed no cardiomegaly; pulmonary vascularity was normal. The electrocardiogram was within normal limits for age. Over the next several days the cyanosis diminished to the point of acyanosis. Physical examination remained unchanged until the time of discharge from the nursery at one week of age, when a short systolic murmur was heard. At two weeks of age the infant was still acyanotic but signs of early heart failure were present. Roentgenograms showed slight cardiomegaly and increased pulmonary vascularity. Electrocardiogram showed increased right axis deviation, and right ventricular hypertrophy. The infant was started on digoxin and diuretics, and arrangements were made for air transfer to the continental United States. Enroute the infant's condition deteriorated and mild cyanosis became evident.

Consider, and rule out, respiratory causes for cyanosis.

Note that significant cyanotic heart disease can occur with initial studies within normal limits, including the physical exam of the heart.

Early signs of failure include tachypnea, tachycardia, hepatomegaly, and cardiomegaly in order of decreasing frequency.

Air transfer is fast, but may be hazardous. A physician should be in attendance.

On admission at Letterman General Hospital the child presented with cyanosis and moderate respiratory distress. All pulses were present and equal. There was moderate hepatosplenomegaly, a right ventricular heave, and a parasternal thrill. On auscultation the second heart sound was single and loud. There was a soft 2/6 systolic ejection murmur in the pulmonic area. Roentgenograms and electrocardiogram were as noted previously. The precatheterization impression was transposition of the

A loud second sound strongly suggests pulmonary hypertension.

great vessels with an associated ventricular septal defect or truncus arteriosus.

See TABLE 1.

At emergency catheterization double outlet right ventricle with an associated ventricular septal defect was diagnosed. Pulmonary to systemic blood flow ratio was 7:1. The infant's condition remained poor and his heart failure was moderately severe in spite of adequate digitalization and furosemide diuretic. Five days after admission, it was decided that he would benefit from banding of the pulmonary artery. Following this procedure, heart size diminished. The infant has remained out of failure for several months now, and has slowly gained weight. There is mild cyanosis at rest. No immediate further surgery is anticipated.

This is palliative, closed-heart surgery, to decrease pulmonary blood flow.

COMMENT

The appearance of cardiac cyanosis in a newborn without murmurs usually suggests transposition of the great vessels with an intact septum. An apparently normal roentgenogram and electrocardiogram support this. When medical facilities are available, this is an indication for immediate cardiac catheterization to establish an accurate diagnosis. One third of infants born with congenital heart disease will expire in the first month of life and the majority of these deaths occur in the first week. Life-saving procedures, such as the balloon septostomy to create an atrial septal defect, can be performed at the time of catheterization.

The fact that this child survived the first two weeks of life indicated that a defect was present which allowed adequate mixing of systemic and pulmonary blood such that cyanosis was not frankly obvious. When the child presented in heart failure, investigation became imperative.

The syndrome of double outlet right ventricle is one of the rare causes of cyanotic heart disease (three percent in most series). On examination, cyanosis is usually not evident. Breathlessness, however, is a prominent feature and causes

frequent feeding difficulties. Noncardiac anomalies are seen in a significant number of patients with the syndrome.

Although both the aorta and pulmonary artery arise from the right ventricle, the atrioventricular valves empty into their respective ventricles. The left ventricle must eject blood through a ventricular septal defect into the right ventricle to maintain the systemic and pulmonary circulations. With an infracristal ventricular septal defect, there is preferential streaming of blood so that left ventricle blood enters the aorta (type I). Type II, the so-called Taussig-Bing deformity, is associated with a supracristal ventricular septal defect, where oxygenated blood preferentially enters the pulmonary artery and saturation in this vessel is greater than in the aorta.

Other associated cardiac defects may be present, such as, aortic stenosis, coarctation of the aorta, and pulmonic stenosis. Angiography demonstrates both vessels arising from the right ventricle with both aortic and pulmonic valves at the same level.

Prognosis is poor, especially if associated with other cardiac or noncardiac malformations. Banding of the pulmonary artery was performed in an attempt to control heart failure and protect the lungs from excessive pressure and blood flow. Those who survive to older ages may undergo correction, whereby a baffle directs blood from the left ventricle, through the ventricular septal defect, and out the aorta, without mixing.

confirmed. The infant was returned to the ward and it expired several hours later.

An accurate diagnosis and prognosis can be made and appropriate action taken.

COMMENT

The major cardiac causes of cyanosis in the newborn include (1) transposition of the great vessels, (2) pulmonary atresia with an intact septum, (3) tricuspid atresia, (4) tetralogy of Fallot, (5) total anomalous pulmonary venous drainage, and (6) aortic atresia (one of the hypoplastic left heart syndromes). The commonest cardiac causes for congestive heart failure in the first week of life include (1) hypoplastic left heart syndrome, (2) transposition of the great vessels, and (3) preductal coarctation of the aorta.

Aortic atresia represents a major disturbance of hemodynamics not only in the infant but also in the fetus. Venous blood enters the right atrium and then passes normally into the right ventricle, and out the pulmonary artery. The left ventricle is hypoplastic or absent, and the aortic valve is atretic. Blood returning from the lungs to the left atrium, passes to the right atrium through a dilated foramen ovale or an atrial septal defect. The descending aorta is filled through a large patent ductus arteriosus. The hypoplastic ascending aorta and arch are filled in a retrograde fashion from the patent ductus. Thus, the right ventricle is the only functional ventricle and it supplies the pulmonary as well as the systemic circuits.

The lungs are subject to blood flow at systemic pressures. If the shunt at the atrial level is small, there is pulmonary venous congestion. The right ventricle works under a pressure and volume overload, and heart failure soon ensues. As failure increases, pulmonary edema increases and cyanosis becomes evident. The combination of failing cardiac output and progressive arterial desaturation leads to progressive, unresponsive acidosis and myocardial ischemia.

In general, aortic atresia is incompatible with life and most infants expire in the first 72 hours of postnatal environment. Survival for several weeks has been reported. Because of this extremely poor prognosis, it is critical that this malformation be differentiated from other potentially salvageable defects.

The diagnosis of aortic atresia is strongly suggested by diminished or absent pulses in all extremities, while a diagnosis of coarctation is suggested by selective absence of the femoral pulses. However, all pulses may be diminished when heart failure occurs in any heart disease. In excluding other diagnoses, the presence of a single second heart sound is evidence against total anomalous pulmonary venous drainage where the second heart sound is usually split. Tetralogy of Fallot and tricuspid atresia rarely present with heart failure; additionally, the roentgenogram in these two disorders along with pulmonary atresia shows diminished vascularity.

The typical roentgenogram in aortic atresia demonstrates marked cardiomegaly, a prominent main pulmonary artery segment, increased vascularity, and often pulmonary venous congestion. The electrocardiogram shows right axis deviation in excess of 140 degrees, and right ventricular hypertrophy. Thus the diagnosis can frequently be made clinically before catheterization.

CASE IV

Congestive heart failure
Left ventricular hypertrophy
Pulmonary vascularity, normal
Cardiomegaly, moderate
Murmur, regurgitant

Diagnosis: Endocardial fibroelastosis

A three-month old infant had an immediate postnatal course complicated by several seizures. Evaluation, however, failed to reveal an etiology for these convulsions. He was discharged on phenobarbital and was followed as an outpatient. At eight weeks of age a pansystolic murmur was heard for the first time from the lower left sternal border to the apex. The second heart sound was normal. Roentgenograms and electrocardiogram were within normal limits but the left precordial leads suggested more left ventricular forces than expected for an infant this age. Over the next two weeks, the infant had two admissions for symptoms of tachypnea and respiratory distress which were diagnosed as bronchiolitis. At eleven weeks of age, the infant was admitted in congestive heart failure. The infant was digitalized and was transferred from the hospital at Fort Ord to Letterman General Hospital.

This is the age when the murmur of a ventricular septal defect typically appears.

Consider, and rule out, vascular ring anomalies, pulmonary edema, and respiratory problems associated with increased pulmonary blood flow.

Physical examination on admission revealed no cyanosis or clubbing and all pulses were normal. Blood pressures in the extremities were normal. On palpation an apical tap was felt. The liver was four centimeters below the right costal margin. On auscultation the first and second heart sounds were normal. There was a loud third heart sound. An apical pansystolic murmur of mitral incompetence was heard which radiated to the back and left axilla. Chest roentgenogram revealed moderate cardiomegaly suggestive of left ventricular enlargement. Pulmonary vascularity was normal. The left atrium was enlarged on the lateral. Electrocardiogram showed left atrial hypertrophy with notched P waves and late inversion in V_1.

Consider the causes of mitral incompetence at this age:
(1) endocardial fibroelastosis
(2) myocarditis
(3) anomalous origin of left coronary artery from pulmonary artery

There was left ventricular hypertrophy with RV_6 of 45 millivolts and pathologic Q waves of 5 millivolts in V_6. T waves were diaphasic in the left precordial leads. The clinical impression was endocardial fibroelastosis.

The patient's condition improved with digoxin and diuretics, and heart failure receded. Subsequently at catheterization, angiography demonstrated an enlarged, dilated, poorly contractile left ventricle, which slowly emptied itself of dye. There was moderate incompetence into a large left atrium. Left atrial pressures were mildly elevated. An aortic root injection demonstrated normal origin of both coronary arteries. The patient was subsequently discharged on maintenance digoxin and diuretics, under close observation. The findings were typical of endocardial fibroelastosis, but myocarditis could not be excluded. One week after discharge the infant expired suddenly while asleep. Autopsy revealed the typical findings of endocardial fibroelastosis.

(4) severe aortic stenosis
(5) endocardial cushion defect
(6) glycogen storage diseases
(7) familial myocardiopathies
(8) congenital mitral incompetence
(9) rhabdomyoma

The response to this therapy is more dramatic in the older infant. Likewise the catheterization can be postponed until the patient is in stable cardiac condition.

COMMENT

Twenty percent of infants and children with organic heart disease develop cardiac failure at some time; 90 percent of those do so in the first year of life. As expected, those with cyanotic heart disease most frequently develop failure in the first weeks of life. By the third month of life, ventricular septal defect, endocardial cushion defects, and endocardial fibroelastosis are the commonest causes of cardiac failure.

The findings of left ventricular enlargement and mitral incompetence at this age suggest one of the cardiomyopathies which were listed in the annotations. A family history of cardiomyopathies or glycogen storage diseases may exist, but in our case it was not present. Isolated mitral incompetence is an exceedingly rare congenital anomaly. Severe aortic

stenosis can be diagnosed on physical examination by considering the nature of the pulse.

Although there may not be specific evidence of myocardial infarction, which would suggest myocarditis or anomalous coronary artery, these diagnoses cannot be ruled out. Acute myocarditis presents with low voltages on the electrocardiogram, but chronic myocarditis occasionally demonstrates left ventricular hypertrophy. Only the diagnosis of anomalous coronary artery is surgically approachable, and the longer the diagnosis is not confirmed, the greater the risk of permanent myocardial damage. For this reason, cardiac catheterization should be performed as soon as the patient is stable.

Primary endocardial fibroelastosis is a disease in which the endocardium of the left ventricle thickens to a glistening white appearance. Rarely is any other chamber involved. The etiology is unknown, but the three main hypotheses suggest that it may be inflammatory, mechanical, or hereditary. Most popular is the belief that it is the sclerotic end-stage of a viral myocarditis. There is a dilated-type, of which our patient is an example, and a rare contracted-type, usually seen in the newborn. The disease is the commonest cause of heart failure in the second six months of life. Murmurs are usually insignificant, unless mitral incompetence occurs as a result of mitral valve ring dilation and deformity. Treatment is symptomatic. The prognosis is generally poor; the majority of patients die in the first two years of life. More recently, an increasing number of survivors until older childhood have been reported, and it is possible that with long-term digitalis, as many as twenty-five percent may survive and actually improve.

TABLE 1

DIAGNOSES TO BE CONSIDERED IN INFANT WITH HEART DISEASE

COMMON	UNCOMMON OR RARE
Right ventricular hypertrophy *with*. . .	
Increased flow	
Mitral and aortic atresia; Coarction syndrome; Transposition of great vessels; Patent ductus arteriosus	Total anomalous pulmonary venous; Double outlet right ventricle, Arteriovenous fistula; Atrial septal defect, including cushion defect
Decreased flow	
Severe pulmonary stenosis and atresia, no ventricular septal defect; Tetralogy of Fallot	Transposition with pulmonary stenosis; Cushion defect with pulmonary stenosis, Congenital tricuspid insufficiency
Normal pulmonary vascular markings	
Respiratory distress syndrome; Infant of diabetic mother; Mild pulmonary stenosis	Arteriovenous fistula
Pulmonary venous obstruction	
Respiratory distress syndrome; Wilson-Mikity syndrome, Total pulmonary venous drainage; Mitral and aortic atresia	Atresia of common pulmonary vein, Cor triatriatum, Supravalvular mitral stenosing ring
Left ventricular hypertrophy *with*. . .	
Increased flow	
. . .	Tricuspid atresia with transposition; Single ventricle without pulmonary stenosis; Double inlet left ventricle, Rare cases of aortic and mitral atresia
Decreased flow	
Tricuspid atresia with pulmonary atresia; Severe pulmonary stenosis or atresia, no ventricular septal defect	. . .
Normal vascularity	
. . .	Endocardial fibroelastosis; Anomalous left coronary artery from pulmonary artery; Severe aortic stenosis, Glycogen storage disease; Rhabdomyoma, Congenital mitral insufficiency; Myocarditis
Combined (left and right) ventricular hypertrophy *with*. . .	
Increased flow	
Transposition of great vessels with ventricular septal defect; Acyanotic tetralogy; Truncus arteriosus; Patent ductus arteriosus, Ventricular septal defect; Coarctation of aorta syndrome	. . .
Decreased flow	
. . .	Pulmonary stenosis/atresia, no ventricular septal defect; Double outlet right ventricle with pulmonary stenosis; Transposition of great vessels with pulmonary stenosis

CONCLUSION

The main purpose of initial diagnosis is to render a decision not necessarily in precise anatomic terms, but as a realistic judgment of the functional severity of the defect. Of all ages of patients, the infant presents the greatest challenge in diagnosis and management.

Cyanosis is a serious sign in the newborn infant. One point has become painfully clear -- after an infant has been in oxygen a few hours, persistent cyanosis should never continue to be observed. Such infants need immediate cardiac catheterization to establish a definite diagnosis and correspondingly accurate prognosis and plan of action.

Congestive heart failure is also a serious sign. In the neonate there should be no delay in proceeding to catheterization. Significant improvement usually will not occur. In the older infant or child, correction of the failure may be tried, and will generally be successful. Catheterization may then be undertaken after the patient's condition has stabilized.

Heart murmurs in the infant frequently have little value in assessing the patient. The infant with serious heart disease may have no murmurs, insignificant murmurs, or nonspecific murmurs (i.e. a loud systolic ejection murmur). Only on occasion will a regurgitant or continuous murmur occur which will lead one to a diagnosis. The nature of the second heart sound will contribute more to a cardiac assessment than any other sound in the cardiac cycle.

Abnormal heart rates have diagnostic value in the two extremes of measurement. Rapid rates in excess of 200 beats per minute should suggest the diagnosis of paroxysmal atrial tachycardia. Unusually slow rates may indicate congenital complete heart block, which may be part of a greater cardiac anomaly (i.e. congenitally corrected transposition of the great vessels with heart block).

In view of the limited response to congenital heart disease, the problem of diagnosis may be approached by dividing the malforamtions on the basis of the electrocardiographic findings, and subdividing on the basis of radiographic findings. Table 1 provides a list of the common and uncommon diagnoses to be considered in the infant with heart disease. Although not the

difinitive answer, it does provide a convenient and an accurate approach to diagnosis.

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A COMPLICATED NEONATE

Management of the Multiple Problems of the Typical Low-birth-weight Infant

CPT Carl D. Robinson, MC, and CPT Stephen L. Done, MC

The intensive care of the sick neonate has become one of the fundamental practices in American pediatrics. Neonatal intensive care units and neonatologists are beginning to flourish throughout the country. The neonate has gained new stature in the human community and "campaigns" on the "promise" that by improving his early care he will function as a better citizen. The case discussed in this paper represents a typical low-birth-weight infant. As the specialty becomes more sophisticated, more of the infant's problems -- which are multiple -- may be managed more effectively.

CASE REPORT

A 765-gram female infant was born of a Gravida I, blood type A positive, serology non-reactive, 19-year-old Caucasian at 27½ week gestation. The onset of labor was spontaneous after pregnancy which was complicated by a partial abruptio involving 30 percent of the placenta. The infant's condition at birth was poor and required immediate resuscitation.

Physical examination revealed an extremely immature female infant with poor heart rate, color and inadequate spontaneous respiration. The ears, eyes, nose and throat were not remarkable except for a 0.5 cm carnivorous hemangioma over the right maxilla, the chest extremely compliant, the heart normal, the external features consistent with a 28-30 week gestation infant. Tone and spontaneous movement were minimal, the Moro was incomplete and the glabellar tap was negative. Gasp, root, and suck were absent. The posturing was full extension.

After the initial resuscitation the infant was stabilized in 45 percent oxygen. Its blood gases were PO_2 = 54 mm Hg, PCO_2 = 57 mm Hg, and pH 7.30. The first day of life was complicated by increasing oxygen need, metabolic and respiratory acidosis, instability of heat balance, hypotension and hypocalcemia. Hypotension was corrected by transfusions of whole blood. Acidosis and hypocalcemia were reversed with sodium bicarbonate and calcium gluconate. The second day was complicated by progressive hyperbilirubinemia, hyperglycemia, glucosuria, acetouria and metabolic acidosis with a stabilizing respiratory status. The persistent hyperglycemia was treated with regular insulin.

By the fourth hospital day, oxygen need was minimal, hypocalcemia and hyperglycemia were no longer a problem. The weight was 680 grams. The infant was receiving maintenance fluids intravenously and was begun on small amounts of oral intake.

By the sixth hospital day in 37 percent oxygen (with PaO_2 = 80-86 mm Hg), the patient began to retain carbon dioxide (CO_2) and became unresponsive to positive pressure ventilation (PPV) administered with ambu bag and mask. The $PaCO_2$ continued to rise and the child became increasingly acidotic.

By the seventh hospital day, CO_2 retention was worse and the infant had began to have apneic episodes. Chest roentgenogram revealed scattered areas of atelectasis. Blood gases progressively deteriorated and the infant was placed on the Bourne's volume controlled positive pressure ventilator with a nasotracheal airway. On controlled ventilation, the respiratory acidosis was corrected and oxygen need subsequently decreased. On the ninth hospital day and second day on respirator, the infant became jaundiced for the second time. By the tenth day the infant's overall condition had worsen. A Gram stain of sputum showed Gram negative rods and the culture yielded Pseudomonas. The patient was started on carbenicillin and later gentamycin. Its status gradually improved. Extubation occurred on the 17th day. No further respiratory problems were subsequently encountered.

Feeding was initially difficult and it was not until the 20th day did she reachieve her birth weight. She continued to gain weight well and by 58 days had doubled her birth weight.

The remainder of her hospital course was without incident and she was discharged to home in good condition at 75 days of age, weighing 2098 grams. Follow-up visits in the clinic reveal a healthy, active growing female infant.

DISCUSSION

We have selected several of the common disorders which occur in the complicated neonate to discuss briefly. They are hypocalcemia, hyperbilirubinemia, "transient diabetes", apnea, infection, and feeding the low-birth-weight infant.

Hypocalcemia

Hypocalcemia in premature infants is a major problem to the pediatrician who cares for neonates. Gittleman et al in a survey of 111 premature infants noted that the serum calcium in 55 of them was less than 8 mg/100 ml at some point during the first week of life. He also noted that the incidence of hypocalcemia varied inversely with the birth weight of the infants and that 75 percent were in the 1080 gm to 1470 gm group. Many reasons for the occurrence of hypocalcemia in the first day of life have been postulated. Among these are (a) the maternal cortisol level, (b) starvation, and (c) functional hypoparathyroidism. The maternal cortisol is elevated at term and cortisol crosses the placenta and at term may be at level that exceed normal adults. Cortisol, it is remembered, opposes parathyroid hormone in calcium and is postulated to cause hypocalcemia by inhibition of bone resorption. Starvation is the second proposed mechanism. Prematures are slow to establish adequate caloric intake and thereby deposition of bone calcium in infants with minimal intake of calcium with abundant undermineralized osteoid could establish hypocalcemia by the same mechanism one finds in cases of rapidly healing rickets. The neonate is functionally hypoparathyroid either due to poor intrauterine stimulation of the parathyroids and kidneys or due to immaturity. Regardless of the etiology,

symptomatic hypocalcemia should be treated vigorously. We employ 200-600 mg/kg as needed of calcium gluconate per dose until asymptomatic, then follow with maintanence doses.

Hyperbilirubinemia

Jaundice is a well-observed phenomenon in both full term and premature infants and is visibly apparent in 50 percent of term and 80 percent of all premature infants. Unique to the newborn period and certain hemolytic and hereditary icteric states, i.e. Crigler-Najhar disease, the jaundice is mostly unconjugated, indirect acting. Unconjugated bilirubin is a highly lipid soluble, non-polar molecule which easily crosses biological membranes and results from metabolism of heme. The life span of the newborn infant's red blood cell is shorter than the adult's. The infant also, perhaps because of the relatively hypoxic intrauterine environment, has an expanded red cell volume which contributes a heavy bilirubin load in the early days of life. Hepatic immaturity, particularly slowness in formation of adequate amounts of glucouronyl transferase, although controversial, is usually given as explanation of icterus neonatorum.

Jaundice may result from many causes: physiological, hemolytic disease (ABO and Rh incompatability, other enzymic and hereditary RBC defects), infection (viral, local bacterial, or generalized septicemia), sequestration of blood in extravascular spaces (e.g. cephalohematomata, bruises), gut hypomobility or obstruction (leading to increased reabsorption of unconjugated bilirubin). Ostructive jaundice although seen in the newborn, will not be considered here.

Unconjugated bilirubin rises to its height on the second to fourth day and returns to normal by the 8th to 14th day in full term infants but tends to appear earlier in preterm infants reaches its apogee later (about 8 days) and returns to normal approximately one week later.

Without question, the premature infant is at greater risk for hyperbilirubinemic toxic encephalopathy, kernicterus. Unconjugated bilirubin is transported in the blood bound to albumin. One mole of albumin will bind two moles of bilirubin -- the first tighter than the second. It is the unbound bilirubin which diffuses into the brain and damages neuronal tissue. The preterm infant is usually hypoproteinemic and thus incapable of binding as great a bilirubin load. Organic compounds and

certain drugs compete for binding sites on albumin. The preterm infant is also more susceptible to conditions favoring kernicterus at lower serum bilirubin levels, i.e. acidosis, hypoxia and asphyxia, hypercapnea, hypoglycemia, and possibly a preexisting neuronal insult.

Diagnosis of the cause of hyperbilirubinemia is important in the management of this problem and therapy should be directed toward correction of the causative situation if possible. Antibiotics appropriate to the infection, exchange transfusion for removal of sensitized red blood cells and bilirubin are specific. Non-specific therapy includes phototherapy usually imploying fluorescent light with wave lengths 300-600 mu usually in the blue spectrum. This promotes photooxidation of the unconjugated bilirubin to water soluble, presumably nontoxic, by-products which are excreted. Although apparently a valuable adjunct to neonatal therapy, the use of phototherapy may have the potential of creating problems for the child later in life. The problems which are directly related to the use of phototherapy have not been sufficiently determined and evaluated. Therefore when one uses phototherapy for the newborn infant who has hyperbilirubemia, he should keep in mind the potential risks involved. Another agent some investigators are employing is phenobarbital in an attempt to induce microsomal enzymic activity. Some use phenobarbital and phototherapy combined. The data at the present time are inconclusive.

Transient Diabetes

"Transient diabetes" in the neonate is a uncommon occurrence. The etiology is at present unknown, but several characteristics are clinically apparent. These include an onset in the first six weeks of life, associated symptoms of failure to thrive. In addition to hyperglycemia, the neonate has dehydration, polyuria, and glucosuria. Many cases have had associated infections. These infants are treated, if detected in time, with insulin and hypotonic saline solutions and caution is taken so as to avoid development of electrolyte imbalance. Long-term prognosis has not yet been assessed.

Apnea

The problems and causes of apnea in the neonate have been summarized by Thibeault. It is necessary to say that the apnea

that occurred in this infant probably is a result of several diverse etiologies among these being chronic hypoventilation, secondary to neonatal atelectasis leading to subsequent carbon dioxide accumulation and hypoxemia requiring assisted ventilation and oxygen. Other major contributing factors are the marked acidosis that occurred, diminished lung compliance, and the immature central nervous system.

Infection

Infection either localized or generalized is a serious problem in the newborn and particularly in the premature infant. This problem has been summarized by Done. The risk to the premature is greatly increased because of immaturity of defense mechanisms, greater likelihood of prolonged rupture of the fetal membranes, greater likelihood of difficulty in labor and delivery, frequent necessity for instrumentation for resuscitation and monitoring in the immediate postnatal period, greater risk of congenital debility, and necessity for prolonged hospitalization. Therapy should be prompt, vigorous, and specific.

Feeding

Of central concern in the care of any newborn and especially the low-birth-weight premature infant is potentiation of continued adequate growth and development. The premature is deprived early of his natural environment and food source, the uterus and placenta. Thus the newborn experiences acute starvation -- the placental supply is cut off. Reversal of the catabolic state and resumption of intrauterine growth rates are dependent upon many factors. Among these are (1) the infant's size, gestational age, and adequacy of nutrition before birth, (2) respiratory capability of infant, infection or congenital debility, (3) thermoregulatory stability and environmental thermoneutrality, (4) level of activity, (5) ability to feed spontaneously and tolerate oral feedings of adequate volume and nutritional value, (6) hormonal factors and maturity of enzyme systems.

Difficulty arises from inadequate knowledge about growth in animals or man or knowledge of nutritional requirements in premature infants. One great obstacle to adequate nutrition in premature infants is the inability to take adequate volumes by mouth to achieve adequate nutrition for growth especially in the first few days of life. Most infants loose weight in

the first few days of life probably as a result of water diuresis, a hypercatabolic state, and diminished intake. Weight loss in premature infants is usually greater and persists longer than the episode for term infants who are "normal" physiologically.

Many approaches to the problem have been used and each has advantages and disadvantages. Among these are (1) early institution of oral feedings, (2) supplementation of oral feedings with regular intravenous fluids, (3) intravenous hyperalimentation, (4) use of formula preparations with higher caloric concentrations (e.g. 0.8-1.3 cal/cc), and (5) use of fats, such as corn oil, or medium chain triglycerides to yield higher calories per volume.

Caloric need. These infants need 120-130 cal/kg/day (sometimes as high as 150 cal/kg/day). Intake must be adequate for resting metabolic needs, physical expenditures, and to restore losses incurred during feedings and growth. Caloric need is intimately related to heat balance. Adequate caloric intake should be achieved as early as possible.

Water. They require 100-150 ml/kg/day. Intake must provide for insensible water loss and obligatory urinary losses secondary to decreased renal concentrating ability.

Protein. The actual need is controversial but is probably 2-4 gm/kg/day. Need is greater in premature than in term infants. High protein diets increase obligatory urinary water loss because of increased solute load and may be responsible for significant metabolic acidosis around 3-4 weeks of age.

Fat. Saturated animal fats are poorly absorbed but unsaturated fats and medium chain triglycerides are well-absorbed.

Carbohydrate. This is the most important source of calories.

Minerals and vitamins. These requirements are suggested in standard texts.

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Probably just as true today –

Remember that inspection is the most valuable of all means employed for physicial examination. Learn to look for abnormalities.

From *Golden Rules of Pediatrics*, John Tahorsky, A.B., M.D.
C.V. Mosby Medical Book Co., St. Louis, 1906.

INFANTILE SPASMS

CPT Bruce A. Epstein, MC

Seizures in infants and children concern the physician who is dedicated to their care. He is cognizant that a significant percentage of persons have some type of convulsions in childhood.* The most insidiously appearing and prognostically poor of the childhood epilepsies is the infantile spasm. Within the past twelve months we have had four patients with infantile spasm admitted to the Pediatric Service, Letterman General Hospital. These represent the clinical spectrum of this disorder and therefore the presentation of the cases with comment and discussion will illustrate the condition, sketch the profile, and present what is considered known and unknown about the etiology, management, and prognosis of infantile spasm.

CASE REPORTS

CASE 1. A $3\frac{1}{2}$-month-old male's admission to Letterman General Hospital was prompted by the abrup onset of episodes consisting of sudden dropping of the head forward one month previously. These occurred in clusters of 5 to 50 times in succession; they seemed to be more frequent in the morning after feeding. The child had his second DPT shot one week prior to the onset of these episodes. Development was within normal range for age and his prenatal and neonatal history was also normal. On examination, there was increased clumsiness, extensor thrusting of the lower extremities, and poor tone on ventral suspension. Laboratory investigations, including metabolic screening, urinary aminoacids, lumbar punctures and skull x-rays, were normal. The electroencephalogram showed hypsarhythmia and the child was started on ACTH therapy. He had a prompt

* A recent study /1/ reported 11.5 percent of 1000 serial admissions in a children's hospital had a history of convulsions, and seizures were the reason for admission of approximately 23 of these patients.

remission in his seizure activity until the prednisone was tapered six weeks post-hospitalization; at that time there was a relapse in the number of spells and the prednisone was restarted. A repeat electro-encephalogram showed a more normal tracing.

COMMENTS: Much has been written to suggest strongly that in certain patients the syndrome of infantile spasms may result from an allergic demyelinating encephalities caused by the pertussis antigen. Critics of this position dismiss this as coincidence because the age of immunization and the age in which the spasms start in most cases is the same. Individual case histories, such as Case I, are suggestive that a relationship does indeed exist between immunization and infantile spasms.

CASE 2. A 7-month-old child, who was delivered by Cesarean section because of fetal distress, required endotracheal intubation immediately because of an Apgar score of 1. Spontaneous respirations were not initiated until eight minutes of age and his hospital course of four weeks was documented by many apneic episodes requiring bag and mask resuscitation. He was admitted to the hospital because of seizures since the age of five months. These were described as follows: his arm jerked forwards and upwards, the head flexed forward, the eyes rolled back into his head and the hips and knees flexed. He had anywhere from 10-20 attacks per bout in succession and as many as five bouts per day. His developmental history was retarded; at seven months he was unable to sit up or roll over; he was hypotonic throughout and assumed a "C-shape" on ventral suspension. Skull x-rays, blood chemistry, urine amino acids, and toxoplasmosis titers were normal. The electroencephalogram was hypsarhythmic. He showed no response to a 10-day course of ACTH therapy and was discharged on Valium® 10 mg, three times a day. Follow-up visit one month later showed no improvement in his development or reduction in the frequency of

his seizures. The Valium® was then increased to 15 mg, three times a day.

COMMENT: It is always difficult for the clinician to assign with certainty a diagnosis of brain damage secondary to anoxia because it is a well-known fact that difficult resuscitations at birth are not always followed later by evidence of cerebral damage. However, in a case where motor milestones were delayed since birth and the child neurologically is grossly retarded, one has enough proof to suggest perinatal anoxia as an etiology for his infantile spasms. His failure to improve with ACTH therapy is also indirect evidence because it appears that children in whom an etiology can be found for the attacks usually respond less favorably to steroid therapy.

CASE 3. A $4\frac{1}{2}$-month-old male infant was hospitalized with a $1\frac{1}{2}$-month history of recurrent seizures. These were described as a sudden flexion of the arms and legs, associated with a cry, occurring in a series of jerks 3 to 4 times per day. He was the product of an apparently normal full-term pregnancy without problems in the initial neonatal period. He developed normally although mother reported that he "startled easily". He held his head up at three months, rolled over at four months, and recognized his mother. Upon admission to the hospital he appeared to be well and in good health. Neurological examination revealed poor head control when raised from the supine to sitting position. On ventral suspension, he assumed a hypotonic posture. Laboratory results were entirely normal except for the electroencephalogram which was interpreted as resembling "hypsarhythmia". The child was started on ACTH therapy and discharged on oral prednisone.

COMMENT: This is the type of candidate who responds well to ACTH therapy. This child was developing normally and no cause could be found for his seizures. He presented with the typical flexor spasms and was observed to have "spontaneous" Moro reflexes while on the floor. Unfortunately, he was lost to followup at this hospital when the father was transferred to another area.

CASE 4. Patient was admitted to the pediatric ward with a known diagnosis of infantile spasms since the age of 6 months. This 3-year-old child on examination was functioning at an 8-10 month level and admission was prompted by a recurrence in seizures. ACTH therapy was attempted without success and a change in anticonvulsant medication was begun as an outpatient.

COMMENT: This child exemplifies the poor prognosis for these children with regard to mentality. He was admitted primarily because ACTH therapy had never been attempted although few of the physicians managing the case thought he would respond favorably at this late date.

DISCUSSION

Infantile spasms are divided into three groups depending on their clinical manifestations. The flexor spasm (salaam attack, massive myoclonic seizure) is the commonest form and its description is fairly characteristic. With startling suddenness the child will fling his arms upward and outward with simultaneous flexion of the head and legs. There is usually a sharp cry and the episode may then be repeated in rapid succession. At times, the entire attack may resemble a Moro reflex. In the less common extensor spasm, the legs extend at the hips and the entire body assumes a "spread eagle" appearance. There is also a third variety confined solely to nodding of the head from neck flexion.

The duration of the spasm, regardless of the type, is usually brief. German physicians termed them "Blitzkramph" or lightning spasms. Frequently, a mother will say, "that was an attack just then" but the spell will be over by the time the physician attention is directed to the child.

There is no real agreement as to whether or not consciousness is lost during the spasm and this is understandable considering the youth of most of the patients, the brevity of the spasm, and the associated mental retardation. They seem to occur more commonly following feeding, or prior to sleep, and upon awakening.

The onset of infantile spasms is usually in the first year of life. Livingston /2/ found the peak incidence to be between 3-6 months of age, with eight days of life being the earliest reported case and $5\frac{1}{2}$ years of age the latest. There is no apparent sex predilection.

Brain damage is a prominent feature of the clinical picture, evidence of which is usually present before the onset of the spasms. Gibbs /3/ reported nine percent of all cases with infantile spasms had other types of seizures before the onset of their spasms. Livingston /2/ showed that 86 percent exhibited clinical evidence of brain damage such as developmental retardation and cerebral palsies before the onset of their spasms.

Etiology

Although the etiology is still uncertain, a presumptive cause is usually determined in about 60 percent of cases. Table 1.

TABLE 1
ETIOLOGY OF INFANTILE SPASMS

Perinatal trauma	Congenital cerebral defect
Postnatal trauma	Tuberculous sclerosis
Encephalitis	Hypoglycemic encephalopathy
Phenylketonuria	Postimmunization encephalopathy
Toxoplasmosis	Postinfectious encephalopathy
Cerebral lipidosis	Maple syrup urine disease
Pyridoxine dependency	Cerebral vascular lesions
Central nervous system syphilis	Kernicteris

Of etiological interest is the relationship between infantile spasms and DPT immunizations. The earliest report of a possible relationship came from Baird and Borofsky /4/ who suggested that it was the pertussis immunization that may be the factor involved. This impression was substantiated by Byres and Mool /5/ who collected a group of 15 children who developed severe cerebral symptoms following administration of prophylactic pertussis vaccine, nine of whom developed infantile spasms. Livingston /2/, however, thinks that this relationship is not one of cause and effect but in all probability one of coincidence, because infantile spasms make their appearance at the same time the primary DPT immunizations are given.

Evaluation

The electroencephalographic (EEG) abnormality in children with infantile spasms was first described by Gibbs and Gibbs /6/ in 1952:

> ... very high voltage, random slow waves, and spikes in all cortical areas. The spikes vary from moment to moment in duration and location, at times they seem to come from another focus, or possibly from multiple foci. Occasionally, the spike discharge is generalized but it is never rhythmically repetitive or highly organized... the chaotic appearance of this abnormality gives the impression of nearly total disorganization of cortical voltage regulation...

This bizarre EEG pattern, defined as "hypsarhythmia" by the authors, is found in 80-100 percent of cases with infantile spasms. A normal or nonspecific tracing may occur in one to 16 percent, according to Hess and Neuhaus./7/

All children suspected of having infantile spasms should be hospitalized in an attempt to find an etiology for their seizures. It is important to recognize some of the causes, such as phenylketonuria and hypoglycemia, because both conditions are remediable to therapy. Although other causes are less important to diagnose, prognosis can be more intelligently predicted if a presumptive cause is established. Furthermore, hospitalization enables the physician ample time to work with the family. A suggested evaluation for a child suspected of having infantile spasms is shown in Table 2. Other tests may be added as indicated.

TABLE 2

LABORATORY TESTS. A SUGGESTED LIST FOR EVALUATION OF CHILD WITH INFANTILE SPASMS

Electroencephalogram	Blood sugars
Urinary amino acid chromatogram	Urinary ferric chloride
Lumbar puncture	Toxoplasmosis
Formal developmental testing	Skull x-rays

The most serious handicap in children inflicted with infantile spasms is not the seizures themselves but the associated mental retardation. Livingston /2/ showed that although the spells disappeared completely in untreated cases by the time the child reached 5, over 95 percent of them are severely retarded. Therefore, until recently the major treatment available was the general care and management of the mentally defective child.

Management

In a search for a therapy to prevent this retardation, almost every drug from Dilantin to tetracycline has been tried, either alone or in combination. Although some authors have claimed success with a particular drug, no consistent results have been found with the same drug by other investigators. The most significant breakthrough toward an effective means of controlling the seizures (and, hopefully, of preventing mental decay) was the introduction by Sorel and Dusaucy-Bauloye /8/ of ACTH into the therapeutic armamentarium. They claimed an improvement in 30 percent of cases. Although recent studies /9,10/ have not been able to show the same improvement in mental function, out of them has come a profile of an infant who may be salvageable with early steroid therapy; he is older than six months but less than one year, has had his spasms less than one month, his neurological development has been normal up until the time of diagnosis, and no cause can be found for his attacks. Therefore, prompt recognition and early institution of ACTH therapy may be beneficial in preventing the heretofore dismal prognosis.

ACTH is given at a dose of 20 units of the gel daily in two divided doses for 10 days. If the seizures do not abate, the dose is increased to 30 or 40 units per day. The steroid therapy is stopped if there is no improvement after two to three weeks. If, however, there is improvement, the seizures stop and the electroencephalogram becomes more normal in appearance, then steroids are given for six to eight more weeks and then tapered. Another six to eight week course may then be given if the seizures reoccur following tapering.

The mechanism of steroid action is completely unknown. Bowers and Jeavons /11/ suggest that under hormonal treatment, there is an acceleration of the natural changes that occur with age. Although ACTH is favored by most authors over prednisone, the latter may be justified particularly when long-term treatment is needed since prednisone can be conveniently given at home.

For those children who do not respond to steroids, Valium® in doses upto 40-50 mg/day has proven to be the most reliable drug in supressing the seizure activity. This drug, however, has been a disappointment in preventing the concomitant retardation.

COMMENT

The clinical and encephalographic manifestations of infantile spasms are presented along with four recent cases admitted for investigation at Letterman General Hospital. The mental decay associated with this form of childhood epilepsy remains refractory to all forms of medication and more research is needed to unlock the mystery of this most dismal disease. It has been nearly 130 years since W.J. West published in a letter to Lancet the first clear description of infantile spasms and the accompanying mental retardation. His letter was in the form of an appeal for help since the patient he described was his own son. Despite our further elucidation of the problem, especially by defining the electroencephalographic abnormality, an answer to Doctor West's appeal for help has not been forthcoming.

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—Bloom where you are planted.
—Unknown

REYE'S SYNDROME

CPT Bruce A. Epstein, MC

Acute encephalopathy in children associated with hepatocellular dysfunction was first described as a distinct clinopathological entity by Reye and his colleagues in 1963./1/ They reported a group of twenty-one Australian children who clinically presented with convulsions, coma, hepatomegaly, and hypoglycemia. The disease proved fatal to seventeen. Massive cerebral edema and fatty degeneration of the liver dominated the pathological picture. This illness is now being recognized with increasing frequency in the pediatric age group with approximately 500 cases having now been reported in the medical literature./2/

CASE REPORT

A four-year-old girl was admitted to Letterman General Hospital (LGH) in an unresponsive state. She had been in good health except for a mild spastic cerebral palsy since birth until two weeks before admission when she developed symptoms suggestive of a mild upper respiratory tract infection. Five days later her mother took her to a physician because of her persistent vomiting. An urinalysis showed 18-20 white blood cells (per high power field) and she was started on Gantrisin®. A urine culture showed 1,000 Escherichia coli. She did well and was able to return to kindergarten until the day prior to admission to LGH when she spiked a temperature to 103 F and became sleepy and lethargic. The child was admitted to a small local hospital and given Tigan® suppositories and fluids as tolerated. The following morning she became tachypneic, lapsed into coma and was transferred to LGH.

On admission at LGH she was semicomatose and responded only to pain. Temperature was 99 F, rectally; pulse 120 beats/min and regular; respirations 40/min, deep and labored. Blood pressure was 110/70 mm Hg. The optic discs and fundi appeared normal. The pupils were dilated but reacted normally to light; doll's eye movements were present. The neck was supple and no lymph nodes were palpable. The lungs and heart were normal. The liver was not enlarged and the spleen was not palpated. She responded only to deep painful stimuli and no abnormal reflexes were found. There were no focal or localizing neurological signs. An intravenous cutdown was performed and sodium bicarbonate and 50 percent glucose were given by "push". She was started on intravenous dexamethasone and maintained on a 10 percent glucose infusion. About five hours after admission, coffee ground material was seen coming from her nasal gastric tube and shortly thereafter she was noted to be oozing from previous injection sites. Blood replacement therapy was started but she continued on a downhill course and at 18 hours following admission her pupils became dilated and fixed. Shortly thereafter, her blood pressure was unobtainable and her pulse slowed. While a blood gas sample was being drawn, she had a respiratory arrest and did not respond to the usual resuscitative measures. Her optic discs remained clearly visualized. There was no reported abnormal posturing or seizure activity before death.

Table 1 presents other laboratory findings.

Postmortem Findings:

Positive findings at postmortem examination were limited to the liver, kidneys, and brain. The liver was symmetrically enlarged and revealed capsular and cut surfaces of yellow-orange color. Frozen sections of the liver showed uniform involvement of all parenchymal cells by severe fatty change; all of which stained positively for fat (Oil Red O).

Fat was also seen in small amounts in the tubular cells of both kidneys; the kidneys were otherwise histologically unremarkable. The brain was described as soft and swollen without evidence of herniation. A small acute subdural hematoma was also noted on the right side. Toxicologic studies were performed on liver, brain, and blood for the presence of any known toxin or virus.

TABLE 1

LABORATORY INVESTIGATIONS. Four-year-old girl. Reye's Syndrome

SERUM (mEq/liter)		Other SERUM		PLASMA (mg/100 ml)	
Sodium	140	Alkaline phosphatase	210 K-A units	Glucose	30
Potassium	5.9			Urea nitrogen	40
Chloride	112			Creatinine	2.6
Carbon dioxide	2.4	SGOT	1126 Karmen U	Uric acid	8.9
Bilirubin	0.8	LDH	410 Wacker U		
Salicylates	25	Ammonia	750 µg/100 ml	pH Venous blood	6.9

HEMATOLOGY/ COAGULATION		SPINAL FLUID	
Hematocrit	34% per volume of cells	Red blood cells	2
White blood cells	14,700/cu mm	White blood cells	1 (mononuclear)
Polymorphonuclearcytes	67%	Protein	38 mg/100 ml
Lymphocytes	22%	Glucose	35 mg/100 ml
Monocytes	6%	Glutamine	26.4 mg/100 ml
Bands	5%		
Platelets	350,000/cu mm		
Prothrombin	11% of normal		
Partial thromboplastin time	100% of normal		

URINALYSIS		CULTURES	
Sugar	0	Blood	negative
Protein	0	Urine	negative
Acetone	"large"	Cerebrospinal fluid	negative
White blood cells	2-3/hpf		
Red blood cells	10-15/hpf		
Ferric chloride	positive		

DISCUSSION

None of the clinical or pathological features of Reye's syndrome when considered individually are pathognomonic; however, when treated collectively, they appear to represent a distinct clinical entity.

In the years since attention was first drawn to the syndrome of acute encephalopathy and fatty degeneration of the liver, the disease has been recognized with increasing frequency. Although the exact incidence is unknown, Dodge et al /3/ reported that about five percent of children dying in their hospital fit the pattern described by Reye et al /1/ in the original paper. The disease appears to afflict children between the ages of six months and ten years, with the majority of the patients being below the age of four. There has been only one reported instance of a sibling being affected, and this occurred after an interval of nine months./4/ No apparent sex predilection has been noted. Norman /5/ reported a seasonal incidence with the disease being seen more frequently in the fall and winter months.

Typically, a child recovering from a mild upper respiratory infection develops persistent vomiting progressing rapidly to irritibility, lethargy, convulsions, and coma. Physical examination usually shows a comatose child with loud, irregular breathing, hypotonia, changing decerebrate and decortacate posturing, but no focal neurological signs. Some will have a nonspecific erythematous rash and about one-half will have palpable livers.

Blood samples usually show a mild polymorpholeukocytosis with a metabolic acidosis and acetonuria. Hypoglycemia is commonly found. The cerebral spinal fluid is under normal pressure, free of cells, with a normal protein; the cerebrospinal fluid glucose will usually reflect the low blood sugar. Evidence of hepatic dysfunction is reflected by marked elevation of serum transaminase and ammonia levels without hyperbilirubinemia. Azotemia is often seen and presumably is a result of hepatorenal failure.

The postmortem findings first described by Reye and then in subsequent reports /6-10/ are fairly uniform. The brain exhibits cerebral edema and sometimes anoxic neuronal changes. No histological evidence of encephalitis or meningitis has

been reported. The liver in all cases exhibits massive fatty metamorphosis while being of normal size or only slightly enlarged, a finding that explains the absence of a palpable liver in half the children. Microscopic examination reveals innumerable fat vacuoles in liver cells without predilection for any specific zones. When specially stained for glycogen, little of the substance is found in the liver cells. Fat vacuoles are usually present in the cells of the loop of Henle in the kidney and the proximal convuluted renal tubules. Other evidence of fatty changes are found to a lesser degree in the myocardium and pancreas.

Reye's syndrome should be suspected in any child who, following a nonspecific respiratory infection, develops persistent vomiting and tachypnea, and then acutely lapses in to unconsciousness. It is, of course, of utmost importance to rule out other causes of this clinical picture, such as diabetic ketoacidosis, and drug intoxication by salicylates and other poisons.

Goldschmidt* has proposed certain clinical and laboratory features of Reye's syndrome as an aid to diagnosis. Table 2. A majority of these should be present sometime during the course of the child's illness. The diagnosis can never be made, however, unless the physician maintains a high index of suspicion for this entity.

TABLE 2

CLINICAL AND LABORATORY FEATURES CHARACTERISTIC OF REYE'S SYNDROME*

CLINICAL	LABORATORY
Seizures	Less than 10 WBC/mm^3 in cerebrospinal fluid
Decerebrate posturing	SGOT and SGPT greater than 100 I.U.
Hyperactive deep tendon reflexes	Bilirubin less than 2.0 mg/100 ml
Tachypnea	Blood ammonia greater than 250 µg/100 ml
Hepatomegaly	Liver biopsy showing fatty infiltration
	Blood glucose less than 40 mg/100 ml

*Goldsmidt MM: Reye's syndrome. A proposed cooperative study. Protocol, 1971. From National Institutes of Health.

Treatment

Treatment has thus far been only supportive and has been directed toward protecting the brain from toxins due to hepatic dysfunction. Hypoglycemia is treated with high concentrations of glucose intravenously, and seizures are controlled with Valium® or other anticonvulsants. Since the cause of death is acute cerebral edema, measures directed towards reducing intracranial pressure are used. Fluid restriction, mannitol, and intravenous dexamethasone are recommended in therapeutic doses. Acidosis which may exacerbate the cerebral edema /11/, is vigorously treated with alkali therapy. Whole blood exchange transfusion has been proposed to remove the ammonia and other toxic substances that accumulate in the blood./12/ The hyperammonemia is further treated by sterilizing the gastrointestinal tract with neomycin, this removing a source of ammonia production.

Prognosis

Unfortunately, the available forms of therapy ultimately prove futile in changing the fatal outcome in the majority of children with this illness. In his original paper, Reye et al /1/ reported a fatal outcome in seventeen of the twenty-one children studied. Fifty percent of them died within twenty-four hours after admission. The clinical course is fairly characteristic: vomiting, initially severe and protracted, which subsides, but the child fails to regain consciousness. This, together with irregular respirations, convulsions, and peripheral circulatory failure, dominates the clinical picture until death.

Etiology

The etiology of Reye's syndrome is unknown. Speculation as to a cause has focused on infectious, toxic, and metabolic lines. Bacterial cultures have been uniformly negative and attention toward an infectious etiology has been directed toward a possible viral agent. The syndrome has been described in association with chicken pox /10, 13/ and infectious hepatitis./8/ The study by Bradford and Latham /6/, however, indicated the ability to isolate viruses in only 12 of 47 cases with Reye's syndrome and the organisms cultured were diverse and from different sources. Since most of the cases occurred in the winter months when mild viral illnesses are prevalent, it is difficult to

attribute the syndrome to a viral infection alone.

The rapid downhill clinical course and the evidence of severe liver disease has caused investigators to consider the possibility of ingestions of toxic substances as an etiology. Among the compounds that have been implicated are aspirin /14,15/ aflatoxins derived from Aspergillus flavus /4,16/, and tetracyclines./17/ Although exhaustive studies have failed to reveal any consistent hepatotoxin, interest in the question of contaminated food with fungal toxins has stimulated widespread studies. Products of the Aspergillus flavus can contaminate cereals and in experimental animals has produced a severe fatty metamorphosis of the liver. Investigators have not been, however, able to implicate these toxins in children.

Popper and Schaffner /18/ state that diffuse fatty changes in the liver can be secondary to nutritional, metabolic, and endocrine causes. The only evidence of any abnormality of this nature has been the common but not universal finding of hypoglycemia; otherwise, no pre-existing metabolic or endocrine disorder has been described.

In view of the remarkable consistency both in the clinical course and pathological findings a more specific etiology will ultimately be defined. Perhaps it is an intrinsic metabolic defect that causes an idiosyncratic reaction in susceptible children to certain insults. A viral infection may be needed to trigger the syndrome in a child suffering from an intoxication with some fungal toxins. Perhaps the syndrome can be produced by several different agents. Whatever is postulated as the etiologic agent, it must be able to explain not only why isolated deaths occur in a family (where presumably all the children are exposed to the same environment) but also the relationship between the brain damage, hypoglycemia, acid-base imbalance, fatty liver, and the relentless downhill clinical course.

COMMENT

There is nothing more devastating in all of pediatrics than the sudden unexplained death of a previously well child.

Over the past thirty years we have witnessed the decline and disappearance of a number of previously fatal diseases. Through the efforts of epidemiological studies and basic research, perhaps the mystery surrounding the symptom complex known as Reye's syndrome will be discovered. Only then will we be able to offer these unfortunate children both effective treatment and a better prognosis.

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But whether our efforts are or are not favored by life, let us be able to say, when we come near the great goal: "I have done what I could."

–Louis Pasteur

CONSTRICTIVE PERICARDITIS IN INFANCY

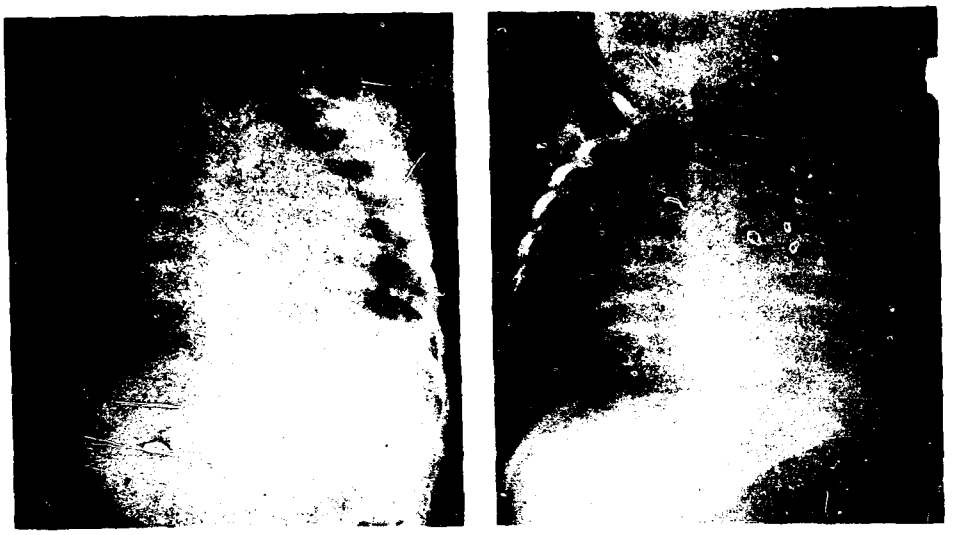
CPT Michael R. Weir, MC

Constrictive pericarditis is known to be uncommon in childhood /1/, and has been described only once in infancy./2/ Purulent pericarditis, according to Gersony and McCracken /3/ has been lethal without pericardiectomy or repeated pericardiocentesis with or without antibiotics. Purulent pericarditis is rarely a precursor of constrictive pericarditis./3/ This paper reports the case of an infant that survived purulent pericarditis that complicated meningitis and pneumonia. He was treated only with antibiotics after a single diagnostic pericardial tap. Subsequently he developed acute constrictive pericarditis that responded to pericardiectomy.

CASE REPORT

A 10½-month-old, oriental male infant, was born in Taiwan to a gravida V, para V mother who died of postpartum hemorrhage, and was adopted by a serviceman and his wife. Pertinent past history included gastroenteritis in June, a febrile seizure in July, and otitis media in September 1970 prior to the onset of the present illness on the third of February 1971. At this time, he had fever and was given clear liquids. Two days later ampicillin was begun orally. The fever persisted, lethargy developed, and a lumbar puncture showed cloudy spinal fluid on 11 February 1971. He was admitted to the Air Force Dispensary (6011) in Taiwan. The white blood cell count revealed 17,500/cu mm with 52 percent lymphocytes and 48 percent neutrophils. Organisms were Gram-positive cocci in chains. Intravenous ampicillin was given and he was transferred to 6217th Air Force Base Dispensary (CCK). Examination at that time revealed an irritable infant with diminished precordial activity, third and fourth heart sounds, absence of murmur and clear lungs. The pulse was suggestive of

pulsus paradoxicus. Kernig's and Brudzinski's signs were negative. Material from the original tap grew out alpha streptococcus, but subsequent cultures were all negative. Massive cardiomegaly and a left pleural effusion were demonstrated radiologically (Fig 1A); fluoroscopy revealed diminished cardiac activity. Electrocardiogram revealed a heart rate of 180/min., axis 90 degrees, and low voltage QRS complexes in the precordial leads. Mercuhydrine® and digoxin were added and ampicillin 150 mg/kg/day was continued. He was transferred to Clark Air Force Base, Phillipines, on 14 February 1971, to have a diagnostic pericardicentesis. The physical examination, at time of arrival, revealed that the third and fourth heart sounds were now absent, Cardiomegaly and opacification of the left lung was demonstrated radiologically (Fig 1B). Lumbar puncture showed five white blood cells of which three were neutrophils and two lymphocytes; the protein was 38 mg/100 ml and glucose was 54 mg/100 ml. Pericardiocentesis on 22 February produced 5.0 ml of blood stained material; the protein was 4.3 gm/100 ml, the sugar 31 mg/100 ml. There were 200,000 red and 52,000 white blood cells per cu mm. Gram stain and cultures proved negative. Air in pericardial cavity and pleural thickening were seen on the roentgenogram (Fig 1C). Cardiac size gradually diminished but clinically he failed to improve. Suspecting constrictive pericarditis, he was transferred to Letterman General Hospital on 4 March 1971. On admission, the temperature was 97F degrees, pulse rate 124 beats/min., respirations 40 breaths/min.; he was irritable, had facial edema, and positive Kernig's and Brudzinski's signs. Examination of the chest revealed diminished breath sounds on the left and egophony on crying without rales or rhonchi. The heart sounds were diminished; third or fourth heart sounds or murmurs were absent. The liver was palpable five centimeters below the right costal margin. The extremities were edematous and pulses were not paradoxical. Admitting laboratory values showed a hematocrit of 41 percent with four nucleated red blood cells/100 white blood cells. There were 17,500 white blood cells per cu mm with 59 percent lymphocytes, 27 percent neutrophils, 13 percent monocytes and one percent eosinophil. The



A. B.

Fig. 1A. Chest roentgenograms. A. Fig. 1B. Taken February 12 and reveals massive cardiomegaly with a globular heart typical of pericardial effusion. Note also the left pleural effusion. B. On February 14 the pericardial effusion was more pronounced in spite of digoxin and diuretics. C. Roentgenogram taken after pericardiocentesis which produced 5.0 ml of fluid. Note the air in the pericardial cavity and pleural thickening at apex. D. Roentgenogram taken five weeks postpericardectomy as part of outpatient follow-up evaluation.

C. D.

sodium was 133 mEq/liter, potassium 5.0 mEq/liter, chloride 80 mEq/liter, and carbon dioxide combining power was 21 mEq/liter. The glucose was 100 mg/100 ml and the blood urea nitrogen was 26 mg/100 ml. The cerebral spinal fluid had 40 red blood cells, 54 mg/100 ml glucose and 16 mg/100 ml protein. The gram stain and culture were negative. Chest roentgenogram showed generalized cardiomegaly and an area of plate-like atelectasis in the right apex. The left pleura appeared thickened. On fluoroscopy the left diaphragm had diminished motion and no cardiac pulsations could be noted. The SGOT was 310 Karmen units, LDH-590 Wacker units, and the alkaline phosphase was 310 International Units/ml. Digoxin was continued and ampicillin stopped.

On 10 March the child had a cardiac catheterization with angiography which revealed a thickened pericardial peel involving all chambers but predominantly the left ventricle; the wedge pressure , right ventricular end-diastolic pressure, and right atrial pressure were 25/16 mm Hg, 27/0-20 mm Hg and 15 mm Hg (mean), respectively. The wave form of the right atrial pressure showed a rapid Y-descent. Marked venous desaturation (PO_2-40 mm Hg) suggested low cardiac output in face of normal arterial saturation (96 percent).

On 12 March 1971, he had a pericardiectomy. At thoracotomy, the heart appeared enlarged with decreased pulsations. Within the pericardial cavity was a large amount of green-cheesy material. This showed large numbers of white blood cells and small numbers of Gram-positive diplococci. No acid-fast bacilli were seen and no organism grew on culture. Following pericardiectomy, the cardiac activity improved associated with increased amplitude on the QRS complex (Fig 2). Postoperative complications were pneumonia on the tenth postoperative day; pneumococcus and Klebsiella were recovered on tracheal aspirate and ampicillin and kanamycin were administered. There was good clinical response. Also, he developed a postoperative wound infection from a cutdown site in the left groin. This responded to local care, as well as an area of cellulitis of the left foot at

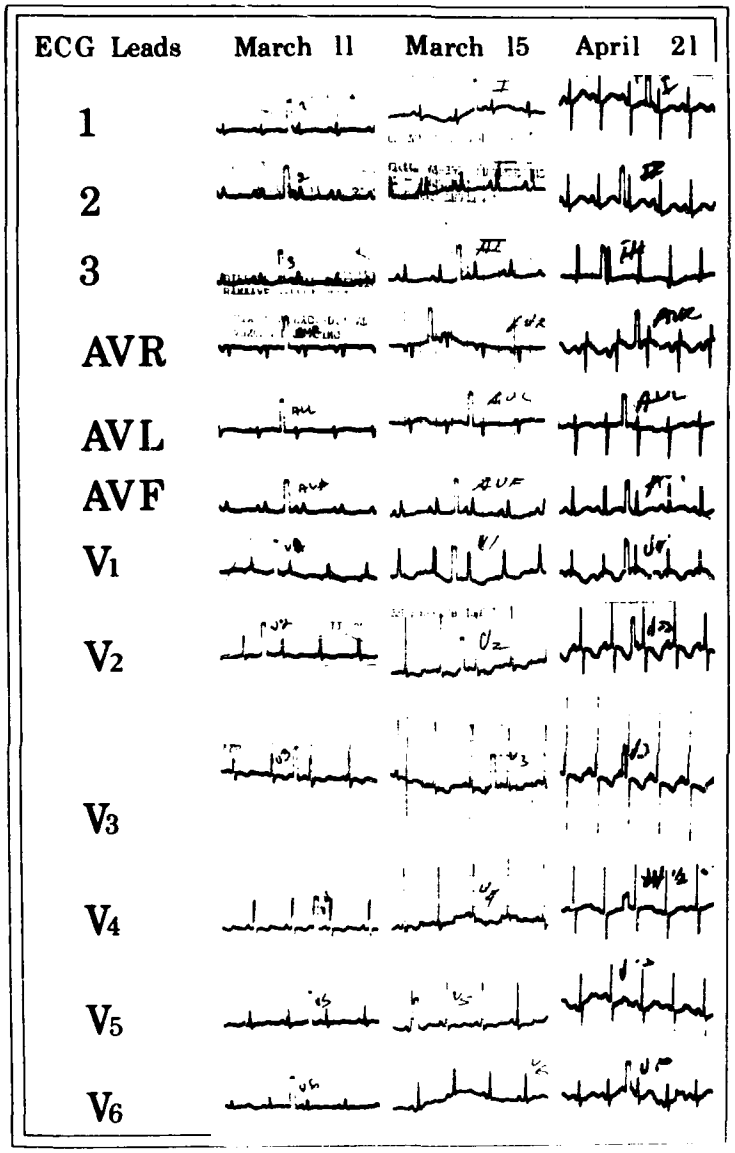


Fig. 2. Electrocardiograms (ECG). ECG, March 11, was taken before pericardectomy and reveals low voltage complexes, right atrial hypertrophy, and nonspecific T waves canges. The ECG, March 15, was taken three days postpericardectomy. Note increase in precrodial leads. The ECG, April 21, five weeks postpericardectomy reveals normal voltages and normal appearing T waves.

at a venipuncture site. He was discharged on 7 April 1971 and was well clinically at followup two weeks after discharge, although pleural thickening was suggested radiologically (Fig 1D).

DISCUSSION

Purulent material in the pericardial cavity in infants is usually a complication of bacterial infection of other sites, commonly the lung or meninges, or both./3/ This is exemplified by our case. The organism may be one of many including Staphylococcus aureus, pneumococcus, Haemophilus influenzae, Streptococcus pyogenes, paracolon bacillus, Neisseria meningitidis, Bacteriodes, Salmonella, or Escherichia coli./3/ Alpha streptococcus, to our knowledge, has not been previously reported. It may have been a contaminant if the organism had been suppressed by the ampicillin. This possibility seems less probable than the selection of this unlikely organism by prior oral ampicillin therapy. Our case, like others, had sterile purulent material in the pericardial cavity, presumably because of the effectiveness of the antibiotics administered for the primary infection.

Gersony and McCracker /3/ described seven infants with purulent pericarditis and reviewed 50 infants in all. The survival rate was improved significantly with drainage, either through repeated pericardicentesis or pericardiotomy. Without drainage, he found no survivors of 21 cases, whereas there were 17 survivors of 29 patients that received drainage. Our case appears to be the only survivor in infancy or childhood without surgical drainage or repeat pericardiocentesis.

Constrictive pericarditis has been described in only one infant, and this was documented by autopsy findings./2/ The exact etiology appears undetermined. In children and in adults, constrictive pericarditis occurs most commonly idiopathically /4,5/ but may occur following tuberculosis, purulent or acute benign pericarditis, or possibly rheumatic heart disease./4/ The development of constrictive pericarditis following purulent pericarditis is uncommon /3,4/, suggesting that either treatment by drainage may actually prevent the development of constriction, or that fatalities occur because constriction develops but is not recognized and treated

surgically. Caddell et al /5/ described a case typical of rapid progression of purulent pericarditis to constrictive pericarditis in a child. Gibbons et al /8/ reported a case, like ours, which also progressed rapidly from effusion to constriction. The fact that the case of Schlesinger et al /2/ and ours both developed a rapid progression to a constrictive process suggests that perhaps infants may be more prone than older patients to acute progression which adds urgency to difficulties as hazards of diagnosis in infancy.

The clinical course suggests that the process was effusive initially before the disappearance of paradoxical pulse. Hancock /7/ emphasizes that there is a significant difference clinically and at catheterization which distinguishes purely constrictive processes from processes in which effusion occurs with or without constriction. He demonstrated that effusion with or without elements of constriction most frequently showed a paradoxical pulse and a predominant systolic descent on right atrial catheterization. If the process is entirely constrictive, i.e. if effusion was not present or was relieved by pericardicentesis, then a third heart sound and a predominant diastolic descent is most frequently noted. He suggested that with use of these clinical signs and laboratory data, the presence of effusion could be evaluated. In its presence, the constrictive element can only be appreciated after the tamponade effect of the effusion is relieved. The disappearance of the paradoxical pulse in our patient coincident with the decrease in size of the heart and clinical deterioration suggests that the effusive component probably disappeared when the constriction developed. The pericardial knock has been associated by Mounsey /8/ with slope of the upswing of the diastolic dip. The pericardial knock is absent when the slope is not great. In spite of the absence of the pericardial knock, the disappearance of the paradoxical pulse is evidence for the evoluation of constriction from effusion.

It is emphasized that drainage is the significant aspect of treatment. The evolution of constrictive pericarditis is postulated to be related to failure to drain the purulent effusion. The distinguishing factors are described thus: (1) effusion is associated with a paradoxical pulse and predominant systolic descent; (2) constriction without effusion is associated with absence of paradoxical pulse, presence of pericardial knock and Kussmaul's sign and a predominant diastolic descent. The development of pericardial

constriction must then be considered with the alteration of the clinical picture.

COMMENT

An infant is reported with a number of unique elements. First, he survived purulent pericarditis without surgical drainage or repeated pericardicentesis. Secondly, he is a one of two infants with constrictive pericarditis and the only infant to survive. Thirdly, the culpable organism appears to have been alpha streptococcus. The development of constrictive pericarditis after a purulent pericardial process is uncommon. The clinical and laboratory data suggest the conversion of an effusive process to a constrictive process.

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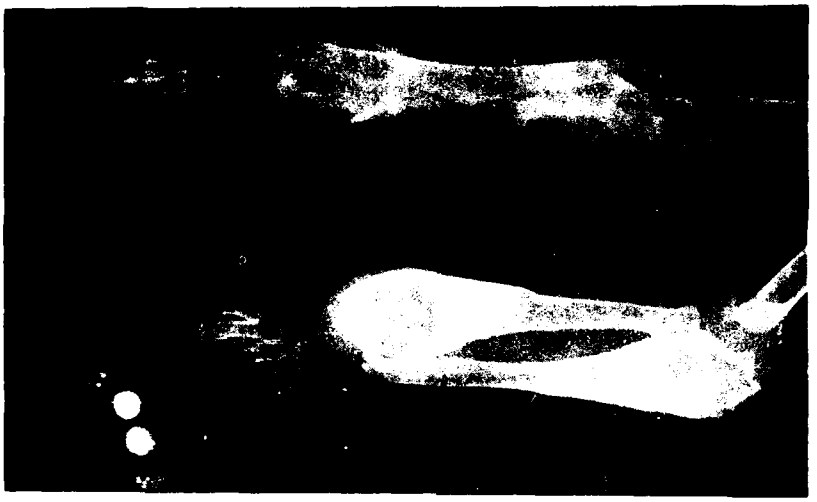

Fig. 1. Cystic destruction of distal radius and ulna.

appeared alert but malnourished and was irritable when examined. The head, eyes, ears, nose and throat revealed no significant abnormalities. The heart had no murmurs and the lungs had bilateral rales but no consolidation. The abdomen was soft without palpable liver or spleen. The extremities showed a non-erythematous, fusiform swelling of the right wrist and swelling of the right and left elbows and left heel. The joints were mobile; movement did not appear to be painful. The swelling was firm but had fluctuant areas. The right wrist had an area of granulation tissue immediately over the most fluctuant area, the site of former purulent drainage.

The admitting chest roentgenogram showed a soft tissue, mediastinal mass that eroded three thoracic vertebrae and the 5th rib posteriorly (Fig 3). The mass had a soft, fluffy appearance at the margin. The abdominal films showed osteolytic lesions of T_{10}, T_{12}, and L_4 (Fig 4A and B). The tuberculin skin test was positive and the gastric aspirate had acid-fast bacilli on smear and culture. Biopsy of the lesion at the wrist showed caseation

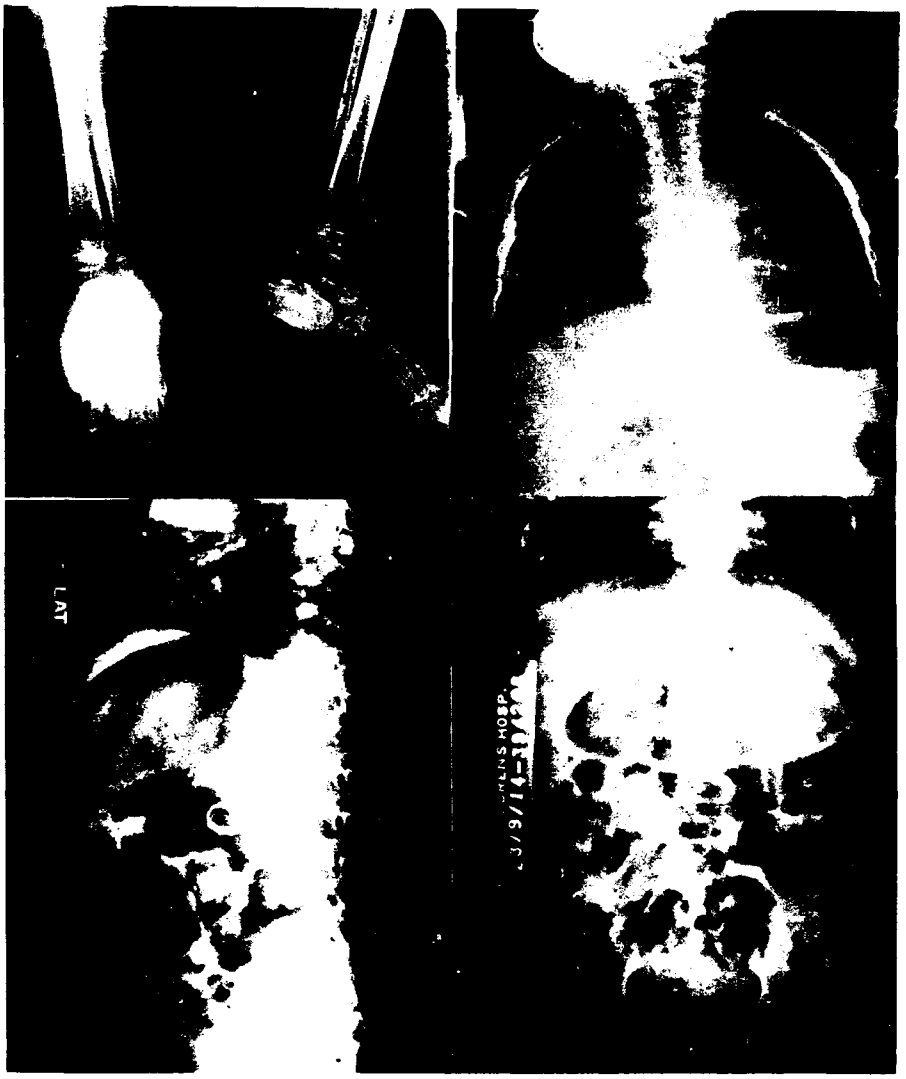

Fig. 2. *(upper left)* Osseous destruction of calcaneı.
Fig. 3. *(upper right)* Roentgenogram of chest, posteroanterior view, which reveals mediastinal mass with erosion of right 5th rib, posteriorly.
Fig. 4. *(lower left and right)* A. Roentgenogram lateral view which shows the destruction of vertebrae, T_{10}, T_{12}, and L_4. B. Roentgenogram, posteroanterior view, which also reveals osseous destruction of T_{10}, T_{12}, and L_4.

with giant cells (Fig 5). The right wrist was splinted in plaster and a body spica applied for vertebral stabilization. Antituberculous therapy with para-aminosalicylic acid (PAS), isonicotinic hydrazide (INH), and streptomycin was initiated. At eight weeks she showed improvement in general condition and appetite.

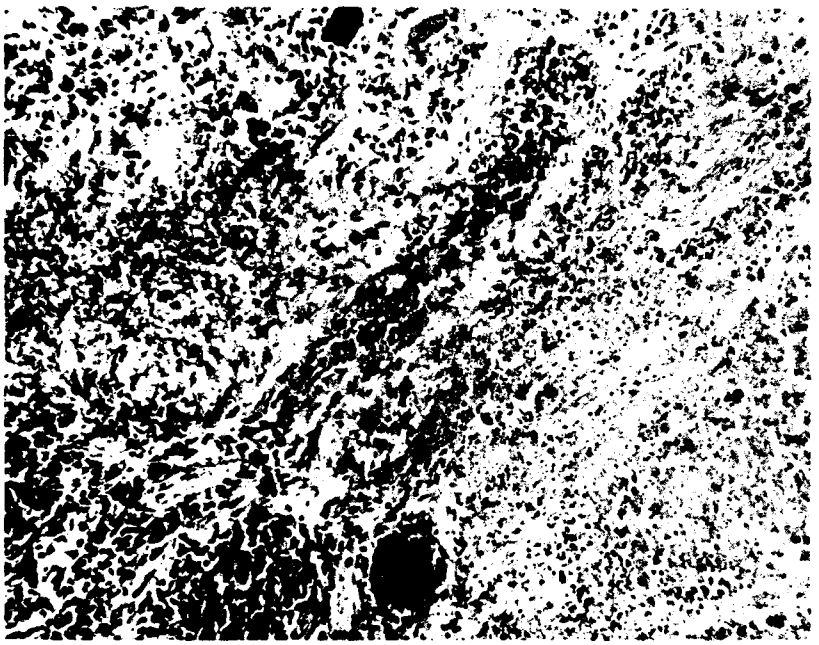

Fig. 5. Biopsy of right radius. Tissue is described as caseating granuloma with giant cells.

DISCUSSION

Osseous tuberculosis in children was found by Lincoln and Sewell /1/ in five percent of children with primary pulmonary tuberculosis. It is a more common complication with generalized tuberculosis where the spread is probably hematogenous. Lymphatic involvement of contiguous invasion may, however, be important in spondylitis.

Of cases with osseous tuberculosis, the most common sites in order of frequency are the spine, the small bones of the hands and feet, the hip, and the knee. Lincoln and Sewell /1/ believed that the functional usage is associated with the site of involvement. Infants that do not walk or stand are less likely to have spondylitis and older children do not commonly have dactylitis. Of the large bones, the site of localization is the metaphysis which is the site of greatest vascularity.

O'Connor et al /2/ reviewed the history of cystic granulomatous lesions of bone. Initially sarcoidosis was not distinguished from tuberculous bone disease but a recognition that it did not have a tuberculous etiology led to the following nomenclature...

—the bone lesions of sarcoid are referred to as osteitis multiplex cystoides sarcoidosa.

—the multiple cyst-like tuberculous lesions of children are called pseudocystic tuberculosis of bone.

—the small cystic lesions typical of tuberculosis in adults are designated disseminated bone tuberculosis.

O'Connor er al /2/ described a case of the third type. O'Malley and Zeft /3/, who also described such a case, further characterized the condition as one of multiple oval lesions aligned with the long axis of the bone.

Clinton-Thomas and Young /4/ described a case similar to ours although no organism was cultured. They proposed that the lesion is distinct from that described as disseminated bone tuberculosis because the bacteremia occurs before the development of the hypersensitivity reaction.

The therapy of osseous tuberculosis involves both antituberculous drugs and careful orthopedic care. The patient's prognosis will probably depend on the extent of involvement of other organs.

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CHILD ABUSE

A Series Reflecting the Profile of Parents and the Child

LTC Henry C. Reister III, MC

In November of 1970 the Letterman Pediatric Service encountered the first of a series of abused children which was destined to number 13 cases and require 390 days of hospital care during a single calendar year. Specific data relating to these cases are presented in the Tables, 1 through 7. For background information on the problem at large the reader may turn to The Maltreated Child /1/ by Vincent J. Fontana. The object of this paper is to present comments upon what has been learned or reinforced in the experiences at Letterman General Hospital since November 1970.

Originally this paper was designed to present a case report of a battered child. As this intent was pursued it became obvious that no one case was "typical" and that going into depth into one case became a "trip" into the subjective. The paper has grown into a compilation of cases which is designed to add to the physician's armamentarium of judgement. Failing to see the material as a whole, or "over-individualizing", seems to be one of the greatest risks to these children. Once a child is protected, individualization is indispensible to rehabilitative efforts.

"Profile" of a Hypothetical Case

Having stated that no one case is "typical", we will draw on the tables to give a "profile" of a hypothetical case which might be seen at Letterman. The child is a white male under one year of age. He was seen in an emergency room last month for a minor traumatic complaint. Now he has a fractured long bone and his mother relates that he must have fallen off the sofa. Mother is 21 and a high school graduate. She is depressed about her husband's low pay, her crowded government quarters, lack of friends, and lack of recreation. The chances are about 50/50 that she has a health problem of

her own, possibly a seizure disorder. The baby was born at term, but there is a better than 50 percent chance that there was minor or major maternal complication of the delivery and an extended nursery stay. Little information is volunteered; mother is quiet, evoking the physician's sympathy. Father is 22 years old, did not finish high school, joined the service, and is now an E-4. The chances are about 50/50 that he is unhappy with his job, his pay, his quarters, and his prospects. His bills for "necessities", as he sees them, exceed his income and he has generally either gone into debt or "moon-lighted" for extra funds. Much of his life is threatening to his survival or his self-esteem. Now so are the physician's questions. The father thinks, "Why don't they just fix the leg and be done with it?" He seems to vascilate between hostility and bewilderment, evoking some sympathy from the physician but also provoking some fear.

LETTERMAN EXPERIENCE

In each of the summarized cases the physician recognized that the trauma observed was in excess of the degree to be expected from the history given. Suspicion was born. From this point on in our cases there was a wide range of activity. Whenever the suspicion was of an undeniable degree the parents were immediately told about it and their voluntary cooperation in hospitalization for protection was solicited and uniformly given. When the suspicion was great but the parents judged unreceptive, hospitalization for the child was achieved on the strength of the need to treat the presenting trauma or to do some laboratory investigation. In several instances admission for care of the trauma preceded the dawn of suspicion and the possibility of battering was raised as a question by residents or staff.

Once admitted or identified as a case of suspected battering, each child and parent became the subject of extensive evaluation at Letterman, with the exception of case 5 who was immediately transferred to Juvenile Court. Cases were evaluated by Pediatric Service, Social Work Service, Army Health Nurse (home visits), Psychology Service, and Child Psychiatry Service. These services worked independently first, and then met in joint session. This multidiscipline approach provided for the deliberation of the facts, and

ventilation of feelings of each contributing discipline. A recommendation about reporting the case and the followup was forwarded to the Chief of Pediatrics. At subsequent regular meetings of this "Infant-Child Protective Council" the cases were periodically reviewed and up-dated.

As the year progressed an initial tendency to disbelieve that parents could do such things to their children gave way to a realization that not only "could" they, they did. Young physicians tended at first to identify with the parents almost to the exclusion of their own common sense, but they ended the year less naïve. There was little tendency to think of the parents as "bad"; possibly because of the extensive literature reflecting abuse as a response to their own pain or possibly because of the early and extensive application of our multidisciplinary approach. Discussion of this general subject throughout the hospital, however, revealed a surprising abundance of outraged "righteousness" among health workers of all types.

From the first case on there was considerable soul-searching, individual and corporate. Fear of "outraged righteousness", evident even in our colleagues, and represented in more tangible form by the police and the courts, was, and continues to be a spectre. Despite our reluctance, however, we were able to bring 10 of 13 cases to the attention of agencies which provide follow-up services. Fears of an avenging force were groundless in so far as the civilian agencies were concerned. We found that police and juvenile authorities both were wedded to the concept of rehabilitation. The single case that remained in military channels was delayed five months in preparations for a General Court Martial. This Court Martial was not held, after a pre-trial sanity hearing undercut likelihood of a guilty finding. In this case we did not offer support and guidance to the man's unit commander who had to make the decision on prosecution. Nonetheless, we must conclude that in San Francisco, at this time, it appears that referral to the civilian police is the child and parent's best assurance of being handled by an authority devoted to rehabilitation. The police are exquisitely sensitive to the recommendations of the Juvenile Court probation officers. Convictions, however, are hard to gain in this type of case because it is impossible to prove who battered the child without either a confession or an accusation by a mate, although it can be easily proved that the child was battered.

It would be gratifying to speak positive of results -- to talk of "cures" and restored families; I can only present the picture as it appears to our staff. We can be gratified that there were no known deaths in our series. Furthermore, it would be fair to say that none of the families is the worse off for our intervention. For those who were professionally involved, it was an opportunity for growth which was, in the main, suitably seized. Only four of the cases are still being followed at Letterman; three of these in conjunction with civil authorities. It is too early to make much positive assertion about these cases. One child (case 2) has just come home from a year of foster care. This case "looks good", but time will tell. On the negative side, in three instances of our 13 cases, subjective material dominated thought, and contact with civilian agencies was avoided. In each of these cases physicians felt that they could depend upon their rapport with the parents to assure followup and safeguard the interests of the children. Of these, two have fallen into that awful category: "LOST TO FOLLOWUP".

COMMENT

No physician is likely to be able to afford to rely on his personal charm or skill to guarantee the safety of a child suspected by him of being battered. This is the conclusion of the law and it is supported by the facts locally. The hospital, taken corporately, has the same weaknesses as its physicians, and should conclude that it can not afford to try to safeguard children by reliance on its own fund of skill and prestige. In the military context, especially, such reliance is folly because the movement of both patients and health professionals is brisk.

Impressive in these cases has been the value of HONESTY. In no instance was there any observed value to subterfuge, half-truth, or euphemism. Parents finding themselves in these situations appreciate honest evaluation of the facts, honest appraisals of future possibilities, and frank information on what the professional plan is. In short, they are in need of someone they can trust. As soon as the physician departs from a truthful course, his swerving is detected, and his

effectiveness is sharply reduced. Dishonesty in dealing with the facts bespeaks the risk of dishonest handling of any other possible matters, for example, the feelings, worries, and revelations the parents may wish or need to make known.

Of similar value has been real compassion. Nothing but honesty should rank above this in terms of giving hope of success. The cases still in followup are responsive mostly to the quality of concern and understanding that is being conveyed by the health professionals involved, in at least two cases workers who were compassionate fared poorly with parents because they were indirect. Simple direct honesty remains paramount.

By looking at all of the cases it becomes evident that we are discussing a social disease which rests on a broad base and must be attacked at each of its roots. Many of the parents themselves were abused or unhappy, and our protective efforts with their children are an approach to this root of the problem. Efforts with the parents after-the-fact are vital to individual rehabilitation but, in terms of the overall problem of child abuse, are analogous to treating a smallpox epidemic with band-aids. Solutions are required for those root factors which are evident in all cases -- more effective education, more satisfying job opportunities, adequate remuneration, better medical care for mothers, better fiscal management by couples, better housing, identification with society in purpose and in recreation, more self-esteem, less isolation. This sounds like an invitation to recapture all of the evils which fled from Pandora's box. And, in fact, it is. We can be sure that until we do, the problem of child abuse will be always with us. While it is with us, we must turn to that one thing left to us by Pandora, namely, hope -- HOPE to lend the strength to approach a seemingly unattainable goal. The answers to the problem of Child Abuse lie in the realm of social medicine and social action. Pediatricians and their colleagues may provide counsel, but it takes an enlightened society to lessen the problem of child abuse.

TABLE 1

BATTERED CHILDREN ADMITTED TO LETTERMAN GENERAL HOSPITAL, 21 NOV 1970 TO 20 NOV 1971

CASE NO	AGE	RACE	SEX	CONDITION FOR WHICH ADMITTED	HOSPITAL DAYS
1	8 Mo	W	F	Fractured Femur	112
2	3 Yr	B	M	Ref. fr. Europe, Nearly Drowned	85
3	7 Mo	W	F	Fractured Skull, Cigarette Burns	47
4	5 Mo	W	M	Fractured Humerus	10
5	13 Mo	B	F	Bruises and Lacerations, Facial	1
6	3 Wk	W	M	Fractured Humerus	23
6	2 Mo	W	M	Fractured Skull, Cerebral Edema	33
7	2 Mo	W	M	Failure to Thrive, Deprivation	4
7	4 Mo	W	M	Failure to Thrive, Deprivation	14
8	5 Mo	W	M	Fractured Skull	42
9	1 Mo	W	M	Fractured Skull	2
10	4 Yr	W	M	Bruises, Belt Marks	7
11	7 Yr	W	M	Social Retardation, Abandonment	7
12	13 Mo	W	M	Bruises, Human Bites	0
13	7 Mo	W	F	Fractured Femur	3
SUMMARY	5/6 Mo	11 W	9 M	10 Fractures	390 Hospital Days
	5 7-13 Mo	2 B	4 F	2 Bruises	365 Calendar Days
	5/13 Mo			1 Failure to Thrive	

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TABLE 2

PARENTS AND SIBLINGS OF BATTERED CHILDREN AT LETTERMAN GENERAL HOSPITAL

CASE No	FATHER'S Age	Branch	Grade	Years Educ	MOTHER'S Age	Years Educ	SIBLING'S Age	Sex	BATTERING PERSON
1	20	USMC	E-1	?	17	?			Mother
2	?	USA	E-6	?	?	?	1 Yr	F	Mother and Father
3	22	USA	E-5	10	20	?	none		Father
4	22	USN	E-5	12	23	14	or		?
5	20	USA	E-2	?	20	?	unknown		Father
6	19	USA	E-2	13	18	12			Mother
7	19	USN	E-2	12	18	12			Mother
8	28	USN	E-6	13	24	12			Mother and Father
9	22	USA	E-5	?	21	?	1 Yr	F	?
							5 Yr	M	
10	26	USA	E-5	?	25	11	3 Yr	F	Mother
11	49		–	?	?	?	6 Yr	F	
							5 Yr	M	Mother and Father
							1 Yr	M	
12	?	USN	E-5	?	?	?	3 Yr	F	Sister
13	23	USN	E-4	9	21	12			?
SUMMARY	Av=24½ Med=22	6 USA 5 USN 1 USMC 1 Fraud	Av=E-4	Av-11.5	Av=21 Med=21	Av=12.3			4=Mothers only 2=Father only 3= Both parents 1=Sibling 3 Unknown

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TABLE 3

PARENTAL STRESS FACTORS

CASE NO	MARITAL FACTORS	FINANCIAL FACTORS	HOUSING	CAREER FACTORS
1	Conceived OOW - Husband not the father	Marked outstanding debts	GOV	Husband happy except for pay
2	Conceived OOW - Born OOW Husband not the father	Poor money management	GOV	Husband unhappy with career shift
3	Husband not the father	Poor money management	GOV	Husband unhappy with assignment outside of his MOS
4	Husband often on watch or on sea duty	"	GOV	"
5	Conceived OOW - Born OOW	"	CIV	Father unhappy in Army
6	Conceived OOW - Baby necessitated marriage	Had to live with maternal in-laws	GOV	Father left college to join Army because of the marriage
7	Father often at sea	Shared an apartment	GOV	Mother unhappy with lot, and working as beautician
8	Marked sexual problems	Poor money management	GOV	Father moon-lighting, but happy with his USN job except for pay
9	"	Insufficient funds	GOV	Father near ETS - no funds for job-hunting trip home, unhappy
10	Living with paramour	On relief and using food stamps	CIV	"
11	Parents separated - Father living with mistress	"	CIV	"
12	" - Occasional sea duty	Mother works "Go-Go"	GOV	"
13	Occasional sea duty	Father moon-lights to make ends meet	GOV	Father awaiting reassignment, not happy with current work
SUMMARY	4/13 conc. OOW 2/13 born OOW 4/13 father not the husband	Financial Stress in Common to All Cases Known	10 GOV 3 CIV	Displeasure Mostly Related to Pay

Abbreviations OOW = out of wedlock MOS = military occupational specialty ETS = estimated time of separation (from service)

TABLE 4

HEALTH FACTORS

CASE NO	BIRTH WEIGHT lb oz	PERINATAL COMPLICATIONS	CURRENT PARENTAL HEALTH PROBLEMS	PARENTS' FAMILIES OF ORIGIN
1	7-11	C-Section due to CPD	Mother "Epilepsy"	Mother's "disrupted"
2	7-0	C-Section due to Breech	Father "Depressed", in therapy prior the battering	"
3	?	Maternal edema and hemorrhage	Father "War Neurosis", "depressed" Father petty crime record (civilian)	Father was battered
4	9-2	none	"	Father reared his siblings
5	7-0	none	Father petty crime record (military)	"
6	6-2	none	Mother "Epilepsy" - Father petty crime record (civilian)	"
7	6-15	Maternal edema and hypertension	Mother "Depressed", "Hypoglycemic", with "Fainting Spells"	Mother ran away from home
8	?	Maternal seizures and pulmonary embolism	Mother "Epilepsy"	"
9	7-4	7 days hosp stay due to maternal minor surgery	Mother "Depression" - "Anemia"	Mother was rejected, considered suicide
10	7-11	none	Mother "Conversion Reaction"	Mother reared her siblings
11	8-0	"Blue Baby" - Tetralogy	Father "Diabetes", given to overuse of alcohol	"
12	"	"	"	"
13	7-11	none	Mother "Epilepsy"	Father's abusive - Mother reared her siblings

SUMMARY

No premature or small for gestational age infants

6/13 maternal complications
1/13 infant complications
5/13 extended infant nursery stay

4/13 Mothers with "Epilepsy"

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TABLE 5

INJURY HISTORY AND "HISTORY" STATEMENTS

CASE NO	PREVIOUS EPISODES Nature	Time P.T.A.	ADMISSION TO LGH Injury	Originally Stated Cause of Injury
1	Bruises, Black eyes	60 days	Fractured Femur	"must have caught leg in crib side"
2	Multiple	variable	Near drowning	"left in bath tub"
3	Rib Fractures	30 days	Skull Fracture Cigarette Burns	"rolled off sofa" "touched the heater"
4	Tibia Fractures	60 days	Fractured Humerus	"car had to stop suddenly, infant thrown against straps of, but not out of, car seat"
5	Humerus Fracture	90 days	Facial bruises and lacerations	"left with father who was drunk"
6	None		Fractured Humerus	"caught arm in crib side while being lifted"
6	(see six above)	60 days	Fractured Skull	"I don't know how it happened"
7	Freq. OPC visits	variable	Failure to Thrive	"Spits up everything he is fed"
8	Facial Bruises	14 days	Fractured Skull	"Mother fell while holding child"
9	Periosteal elevation of humerus	est. 14-21 days	Fractured Skull	"rolled off a dresser"
10	?	?	Bruises and Belt marks	"I beat him to save him from a worse beating by ------" (her paramour)
11	Shuffled and abandoned	varying	Abandoned	"tired of him"
12	None		Bruises and Human bites	"left locked in car with sister"
13	None		Fractured Femur	"must have climbed out of her crib"

SUMMARY

At the time of occurrence of the admitting injury, there had been past medical contact in 8/14 episodes.

Abbreviations P.T.A. = prior to admission OPC = outpatient clinic

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TABLE 6

ADMINISTRATIVE - POLICE REPORTS AND ACTIONS

CASE NO	POLICE REPORT TO	POLICE ACTION	PROSECUTION
1	MP to San Mateo County	Investigation	None
2	None	None	None
3	MP to CID	Investigation	General Court Martial proceedings started and abandoned after 5 months.
4	None	None	None
5	MP to San Francisco County	Investigation	None
6	MP to San Francisco County	Investigation	None
7	None	None	None
8	MP to San Francisco County	Investigation	None
9	None	None	None
10	None	None	None
11	None	None	None
12	MP to San Francisco County	Investigation	None
13	None	None	None
SUMMARY	1/13 Military Police only 5/13 via Military Police to civilian police 7/13 no police report	6/13 investigated by police	0/5 cases investigated by civilian police came to prosecution 1/1 cases investigated by military police came to prosecution

Abbreviations MP = Military Police CID = Criminal Investigation Department

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TABLE 7

ADMINISTRATIVE - JUVENILE COURT REPORTS AND ACTIONS

CASE NO.	JUV. CT. REPORT TO	JUVENILE COURT ACTIONS	PRESENT FOLLOW UP BY
1	San Mateo County	Foster care one year	San Mateo County
2	Alameda County	Guardianship for Maternal Grandmother	Alameda County
3	Marin County	Foster care one year	Marin County and LGH
4	None	None	Lost to Followup
5	San Francisco County	Guardianship for Maternal Grandmother	Los Angeles County
6	San Francisco County	Wardship with home care	San Francisco County
6	San Francisco County	Foster care one year (with PGM)	San Francisco County
7	None	None	Lost to Followup
8	San Francisco County	Foster care three months	San Francisco County and LGH
9	San Francisco County	No investigation; (father's enlistment expired; family moved)	Referred to Pennsylvania authorities by San Francisco Co. Juv. Court
10	San Francisco County	None, case recorded for record	LGH
11	San Francisco County	Wardship with home care at father's	Solano County
12	San Francisco County	Wardship with home care	Solano County
13	None	None	LGH
SUMMARY			
	10/13 cases reported to Juvenile Courts	2/10 to foster care with strangers 4/10 to foster care with relatives 3/10 home care with superviston 1/10 recorded only	10/10 reported cases have on-going follow up and supervision 2/3 unreported cases have been lost to follow up

Abbreviations PGM = paternal grandmother LGH = Letterman General Hospital
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Reference

1. Fontana VC: The Maltreated Child. 2nd edition. Springfield, Illinois: Charles C. Thomas, 1971

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Miss Eiko Aoki, secretary to the Chief, Pediatric Service, has typed all the narratives for this symposium, the second prepared by the Pediatric Service this year. The Acknowledgment which appeared on page 104 of this volume, January 1971, can be repeated with sincerity . . .

To her we feel and say "Thank you" for a job well done.

CONGRATULATIONS

As of the first day of January 1972, Pediatric Service becomes a separate professional department at Letterman.

The Editorial Board extends our sincere congratulations to Lieutenant Colonel Stewart and his new Department of Pediatrics. With our congratulations also goes our sincere appreciation for his various contributions as a guest editor of *Present Concepts* over the past three years.